



**PLS Data Base to  
Continue as**

**Memorial to Frank Reyerse** The Website which contains the PLS Database was originally built and maintained by Frank Reyerse who was diagnosed with PLS in the early 90's.

<http://www.geocities.com/freyerse/>

He passed away in November of 2005. He will be sadly missed but his Website, which in the last 10 years was his mission and passion, will live on. Jim and Thurza Campbell will keep the Database updated as a living memorial to Frank. The site has been modified so that Campbell's will receive your form when you sign up or change your data. Frank's Website is a big help to over 600 predominantly PLSers in 24 different countries. Please list yourself, or check your listing so we can update your data. The more information we put out there, the easier it will be for you to find others near you to synapse (connect!).

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## EVENTS

**Event Reports Spring  
Fling, April 1-2** *Reported  
by and masterfully  
orchestrated by Ronnie  
Grove*

We had a great social meeting with lots of visiting, talking, laughing and relaxation. Nothing too serious except what can be more serious than meeting

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

others who know where you're coming from? The climax of the Penny campaign was the highlight of our weekend. In the beginning, there had been doubt and apprehension. There were times I questioned my logic. A few wondered if I were insane. A million pennies! Who, in their right mind, would undertake such a task? (Who says we are in our right mind?) Steve Lockwood placed a 5 gallon water jug in the conference room on Saturday at the Spring Fling. I knew some of those coming were bringing pennies and I had asked friends, family and co-workers locally to bring their collections and dump them on Saturday. I thought the water jug would be a joke. That's a big jug and I expected maybe 2 to 3 inches of coins at the most. Well, that baby was over half full at day's end plus \$50.00 of rolled coins that we didn't drop in, plus a big bag of pennies in my trunk that had gotten overlooked and a coffee can that we emptied the flower vases into! ....The jug had a total of "new" money at \$188.65 plus we picked up \$590.00 in checks on Saturday and a total of \$95.08 in cash from other sources. Bottom line is that as of right now my records show \$16,211.75 and counting. Vaughn paid a tribute to Jeannie Young from NJ who is a Penny Captain and had her diagnosis changed to ALS. She is now bedridden with only a little hand

movement and speaking with the alphabet board and eye movement. Her team made their goal of \$100.00 every month. Just believe in yourself and those around you. We did it! Everything from here on out is gravy. Let's hope we get lots of gravy. Fundraising is an ongoing thing. Ideas are important. Never be afraid to mention yours. After the conventional ways are used up we start to look for more ideas. They won't all work but sometimes an idea can be the beginning of something that does work. It can be a fun thing, a silly thing, or a very serious thing. What does it matter if in the end it produces dollars for research? Keep the ideas coming and keep those penny jars out. Who knows how much we can raise by the time the jars wear out? That's my two cents worth. And THANK YOU, THANK YOU, THANK YOU each and everyone that helped in any way. You are the BEST!!!! Who wants to take on the next project? The Francis Asbury United Methodist Women fixed our meal on Friday. Co- workers from the bank helped serve it and clean up and on Saturday they returned to me a check for \$215.00. I was the topic of the sermon Sunday. We have a new minister and he was not aware of exactly what my group was. He said when he walked in and saw all these happy people on walkers, using canes, and wheel chairs it brought

tears to his eyes. So, actually, all of us were part of that sermon. . . . *and from Sue Me* I got a couple of checks in the mail and one coming yet so you will have a little more to add to your total from me! It was a great thing to see all those people coming in with their pennies and supporting us! The jar Ronnie is talking about was the size of a baby food jar LOL. Us pirates tried to plunder her pennies but she held strong and she came out the winner! I just couldn't compete! Ronnie, you are the BEST!! Thanks for all you

Synapse - Spring 2006 Edition Page 2 to Illinois. From there we will cut across on 70 to D.C. We will post restaurant and hotel connections for key cities where we have lunch and stop for the night. **10<sup>th</sup>** Lunch - Denny's 2830 Lenwood Road Barstow CA 92311 (760) 253-2533---- Bunkdown - Best Western Royal Inn 1111 Pashard St. Needles CA 92363 (760) 326-5660 **11<sup>th</sup>** Lunch - Red Lobster 2500 S. Beulah Blvd. Flagstaff AZ 86001 (928) 556-9604----Bunkdown - Best Western Arizonian Inn 2508 Navajo Boulevard Holbrook, AZ 86025 (928)524-2611 **12<sup>th</sup>** Lunch - Applebee's 1560 W. Maloney Ave. Gallup, NM, 87301(505) 726-0401----Bunkdown - Rio Grande Best Western 1015 Rio Grande Blvd Albuquerque, NM (505) 843-9500 **13<sup>th</sup>** Lunch - Denny's 3403 E Route

have done!

**Upcoming Events Route 66 Ride Across America June 10<sup>th</sup> - June 19<sup>th</sup> of 2006.** 9 days 3000+ miles, and 12 states! Join the Fischer's in their fight against motor neuron disorders by requesting your information packet at [rt66@elegancenm.com](mailto:rt66@elegancenm.com) or by calling 505-885-1289. The Travel Route <http://www.historic66.com/> We will be on Route 66 from California

66 Tucumcari, NM 88401 (505)461-3094 -- --Bunkdown - Best Western Amarillo Inn 1610 Coulter Dr. Amarillo, TX 79106 (806)358-7861 **14<sup>th</sup>** Lunch - Western Sizzlin 2107 S. Main Elk City, OK 73644 (580)243- 2100----Bunkdown - Saddleback Inn Best Western 4300 SW 3rd Street, Oklahoma City OK 73108 405-947- 7000 **15<sup>th</sup>** Lunch - Funddruckers 4329 South Peoria Ave. Tulsa OK (918) 742-1714--- -Bunkdown - Comfort Inn 3400 S Rangeline Rd Joplin, MO 64804(417) 627-0400 **16<sup>th</sup>** Lunch - Shoneys 1015 South Jefferson Avenue Lebanon, MO 65536 (417) 532-2788----Bunkdown - Best Western Inn At Park 4630 Lindell Blvd, St Louis, 63108 (314) 367-7500 **17<sup>th</sup>** Lunch - Outback Steakhouse 3700 U.S. Hwy. 41

S.Terre Haute In (812)  
232-6283---Bunkdown - Best  
Western Indianapolis South 4450  
Southport Crossing Drive  
Indianapolis, Indiana 46237  
(317)888-5588 **18<sup>th</sup>** Lunch - The  
Spagetti Factory 36 W.Fifth St  
Dayton Ohio (937) 461-3913-  
---Bunkdown - Best Western  
Clarmont

Inn 650 S High St Columbus, OH  
43215 (800) 937-8376 **19<sup>th</sup>** Lunch -  
Bob Evans 775 National Road  
Wheeling WV.26003  
(304)232.4139---- Celebration  
Dinner TBA---- Bunkdown - TBA

**Route 66 Ride Update** *Contributed  
by Linda Gentner* I'm not sure how  
many of you have kept up with the  
incredible amount of work that  
Shellie is doing to pull off this ride  
across country. Shellie is dedicating  
the ride in memory/honor of Frank  
Reyerse who was one of the  
pioneers for PLS. After his diagnosis  
changed to ALS he continued with  
his website and the Worldwide PLS  
Registry. He passed away in  
November 2005. He did SO much  
for all of us. The ride will end in  
Washington DC. There will be a

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call Shellie at 505-885-1289 or  
505-302- 8262

**Upcoming SP Connections April  
22, Saturday - Norman, OK** SPF  
Connection event at the Red Lobster

party and celebration dinner. Frank's  
wife, and 3 of his 4 children from  
Canada will be traveling to DC for  
the tribute. Our Board Member, Jean  
Chambers, who also lives in Canada  
will travel with them. I betting there  
won't be a dry eye in the house at  
the Celebration Dinner. *More  
updates* Riders and bike clubs  
already registered include Winged  
Riders of Artesia, American Legion  
Riders of Carlsbad, CMA Wings of  
the Son Riders Carlsbad, Carlsbad  
Radio- Gerry Florez and Mary  
Walterscheid Carlsbad Rotary.  
Early- bird sponsors include State  
Farm Insurance; Winged Riders of  
Artesia; Knights of Columbus  
Carlsbad; Yamaha; Councilman  
Manuel Anaya and Elegance Debby  
Perkins. Joe Vargas brought  
Christopher Coppola to New Mexico  
to try to make Albuquerque little  
Hollywood. Joe is an event  
coordinator. He brings in talent,  
media and has connections to gather  
sponsorship. Over the next few  
weeks he will be working with me to  
make this happen. To register as a  
Rider in any portion of this event or  
to become a Route 66 Sponsor:  
email [rt66@hotmail.com](mailto:rt66@hotmail.com) or

Inn in Norman, Oklahoma from  
12:00 p.m. to 3:00 p.m. Terry Jones,  
an ALS survivor of 33 years is going  
to speak. Contact Mark Dvorak -  
405-447-6085

[czechmarkmhd@yahoo.com](mailto:czechmarkmhd@yahoo.com) **May 21, Sunday - Burlington, MA** SPF

Connection event at the Lahey Medical Clinic in Burlington 2:00 p.m. - 4:00 p.m. Contact Kathi Geisler- 978-256- 2673

[kathi@sp-foundation.org](mailto:kathi@sp-foundation.org) **August 12, Saturday - Southampton, Long**

**Island** Southampton High Class of 1964 60th Birthday Bash/Benefit for the Spastic Paraplegia Foundation(SPF) - 6 p.m.- ? Cocktail Benefit held at my family home in Southampton, Long Island (65 Lewis Street). We are asking for donations, rather than an admission fee.

Contact Carolyn Sartain Anderson - 772- 388-8499

[beachmusic1012@bellsouth.net](mailto:beachmusic1012@bellsouth.net)

**October 6-7, Friday and Saturday -**

The 5<sup>th</sup> "**Autumn in Carolina**".

SAWCAR races on Saturday!

Holiday Inn Select (Madison Park)

\$64.00 plus tax per night. Contact Don Wilson

**TeamWalks 2006 September 16 –**

**Pleasanton, CA** Valley Community Church is partnering with us to hold a 5K Walk and 10K Run along with the TeamWalk. There will be TV coverage, newspaper articles and radio spots announcing the walk and run. All proceeds will go to SPF. Contact Linda Gentner 510-651-5676 [lkgentner@aol.com](mailto:lkgentner@aol.com)

**SP**

## **FOUNDATION**

### **A Letter from the SP President**

Spring has sprung, the cherry blossoms in Washington, DC have hit their peak. But like our disorders there is always a surprise in store, this April morning we actually had snow flurries. But unlike our disorders, the snow did not last long. For the 2006 Research Grant Program, the SPF has budgeted \$400,000. Grants in the range of \$40,000 - \$60,000 per year will be awarded for one and two year proposals. Proposals on any aspect of hereditary spastic paraplegia (HSP) or primary lateral sclerosis (PLS) will be supported. With the 2006 grants, SPF has awarded over one million dollars for medical research. The Board of Directors welcomes Dr. Malin Dollinger and Karen Johnson as our newest members. Dr. Dollinger lives in California and is an accomplished oncologist and author. Karen Johnson is an airline pilot for Northwest Airlines and lives in North Carolina. Please join us in welcoming Malin and Karen to the SPF Board of Directors. Our Annual Board Meeting was held in Nashville, TN on April 1, 2006. As mentioned in the previous Synapse, the BOD is working with the Nonprofit Center in

Nashville. One of the centers consultants, Michelle Conn, joined the meeting in the afternoon to discuss our fundraising efforts. To help move SPF to the next level, it is recommended that we consider hiring a fundraising professional. The BOD will first need to develop a job description and then progress through a search process. Shellie Fischer is busy organizing the Route 66 Ride Across America. If the Ride

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card. The proceeds will be used to offset her expenses for the trip, such as for gas and hotel rooms. The raffle tickets are \$1 each, please contact Shellie

([MUDLUPII@hotmail.com](mailto:MUDLUPII@hotmail.com)) if you are interested in buying tickets.

Plans are being made for the 2006 National Conference and TeamWalk. Betsy Baquet, organizer of the first NY TeamWalk Connection last year, will serve as Chairman for the 2006 weekend. It will be held on Long Island in September, the date will be announced once Betsy secures the hotel and park. The Expanding Horizons program is off and running. The program is designed to form a regional network for community members to meet, talk and support each other. This is especially desired for people who are recently diagnosed with PLS and HSP, they appreciate learning more about these rare disorders. If you haven't joined already, please consider

will be passing near your hometown, please consider cheering the group on at lunch or dinner along the way. To register to host or attend a "Cheer On Connection", contact Kathi Geisler at [Kathi@sp-foundation.org](mailto:Kathi@sp-foundation.org) or 978-256-2673. If you are not able to personally cheer them on, please consider buying raffle tickets from Shellie. The prize is a \$500 WalMart gift

becoming an Ambassador for your state. Please see the Expanding Horizons article for more details. Finally, congratulations to Ronnie Grove, for a successful Saving Pennies For SPF campaign. At the beginning of the campaign, Ronnie set a goal of one million pennies or \$10,000. With the help of Team Captains and their team members, this campaign raised over \$16,000, 60% over the original goal. Great Job! Annette Lockwood

**The SPF has budgeted \$400,000 for its 2006 Research Grant**

**Program** Grants in the range of \$40,000 - \$60,000 per year will be awarded for one and two year proposals. Proposals on any aspect of hereditary spastic paraplegia (HSP) or primary lateral sclerosis (PLS) will be supported. Research grants are offered primarily as "seed monies" to assist investigators with new ideas, those in the early or pilot

phase of their studies, or as additional support for ongoing investigations with demonstration of need. We anticipate that studies funded by the SPF will develop into projects that can successfully attract future funding from other sources. The title of each study funded by the SPF, the name of the principal investigator, as well as his/her institution, city and state will be published on our web page, newsletter, annual report and wherever else the SPF feels is appropriate. Accordingly, each grant application must include a title understandable to the lay public that will be used as the SPF wishes. All other parts of the grant application are considered confidential and will only be released to members of the SPF Scientific Advisory Board, its consultants, and the Board of Directors.

### **Expanding Horizons Program**

Over the past ten years, we have seen the birth and development of HSP and PLS online patient

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Expanding Horizons program to facilitate support and help (a template will be provided). - Individuals will be invited to remain in the regional network to receive future emailings, provide additional contact information and to get involved in support and information efforts. Phase 3: Execute the

communities. Individuals from around the world are helping one another every day, offering critical support and information services via the internet. Now, we'd like to bring that networking, support and help a little closer home through the new Expanding Horizons Program and the help of volunteer State Ambassadors. Here's how the program will work: Phase 1: Establish the network and locations of people - Ambassadors will receive an Excel spreadsheet of contact information for people in their state known to have HSP or PLS. The number of people will range from a few to 150 and the information will range from just names and emails to full mailing addresses. - The spreadsheet will indicate the counties that individuals reside in, so that Ambassadors can use a map to ascertain the general locations of people. Phase 2: Announce the program - Ambassadors will send out an emailing, informing people of our new

program - Ambassadors will receive information of new people in their states who join our community and send out Welcome emails to them. Depending upon the state, this may be 1-12 people over the course of a year. - Let people who live within reasonable proximity to one another know they are "neighbors" and

encourage them to meet. - Send out periodic emails as needed to maintain the regional network. - Other program activities could include: a. Develop state wide resource lists, such as medical practitioners, durable medical goods, advocacy assistance, etc. (a template will be provided) b. Send announcements to the media to raise awareness of SPF conditions c. Hold periodic group support meetings or more formal events d. Develop outreach strategies to locate other people with HSP or PLS Criterion for Ambassadors - Have an SPF disorder (HSP or PLS) or are related to someone who does. - Are people who seek to enhance the quality of life for individuals and bring energy and commitment to the program. - Are comfortable with email, internet searches and Excel and Word applications. - Can make a commitment of one year. - Will serve as a positive ambassador for the Spastic Paraplegia Foundation in attitude and is knowledgeable about SPF initiatives. - Will work as a member of the Ambassador Team. - Will respect and maintain the confidentiality of our members and not share personal information outside of

SPF initiatives. Additionally, Ambassadors will respect the desire of individuals who do not want any further contact. We have ambassadors for the following

states: AZ, CA, CT, FL, GA, IL, IA, IN, ME, MI, NC, ND, NM, NY, OH, OK, PA, SC, TN, TX, VA, WA. Plans are incomplete at this time as to how best to reach those without computers. Please contact Kathi Geisler at [kathigeisler1@aol.com](mailto:kathigeisler1@aol.com) or 978-256-2673 if you'd like to get involved as an Ambassador for the Expanding Horizons program.

### **Documentary to be Produced about PLS /HSP** Elizabeth

Thompson, an Emmy Award-winning documentary filmmaker and Academy Award nominee, learned recently that her mother has PLS. To honor her mother's struggle and bring awareness to PLS and related conditions (HSP), she is creating a documentary. SPF's Medical Advisor John K. Fink, MD, will serve as the film's medical advisor and SPF will assist Elizabeth in research and fundraising. Elizabeth has a special request for individuals with PLS who served in the Gulf War, professional athletes and migrant workers who are afflicted with the disease. Please contact her:

[elizabeth@thompsonfilms.com](mailto:elizabeth@thompsonfilms.com) or 510- 208-5084. Tax deductible donations to support this project can be sent to her fiscal sponsor: Film Arts Foundation, 145 Ninth Street, Ste 101, San Francisco, CA 94103 - 415-552-8760 [www.filmarts.org](http://www.filmarts.org) . Please note "Elizabeth Thompson" on the subject line.

## **Living with HSP/PLS**

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[www.elderhostel.org](http://www.elderhostel.org) Many of our Elderhostel programs can accommodate participants with special needs, particularly our U.S.-based programs. Due to the wide range of mobility or other special needs issues, however, we do not compile a comprehensive list, and recommend that you locate a program, review the program description carefully, and determine if it sounds suitable to your situation. We then ask that you make inquiries with us once you have chosen a program, and to outline for us your specific requirements and mobility and/or other special needs issues. We will be very happy to make inquiries for you with our specific Program Providers who host the programs to confirm if your enjoyment of the program would be compromised in any way. We can then email you the Provider's response. Because we offer more detailed information on our website, we encourage you to visit [www.elderhostel.org](http://www.elderhostel.org) to review the program descriptions as well as the "Detailed Itinerary" link within each description which outline the day-to-day activities. If you have any further questions, please do not

**Elderhostel Carlos Metcalf -  
Customer Service, Elderhostel,  
Inc.**

hesitate to call us, toll-free, at 1-877-426-8056, M-F, 9am-6pm (ET).

### **Physical Therapy Sessions**

Contributed by Kathi Geisler My neuro referred me to have four PT sessions to set up a regimen. The first is the standard stand Achilles stretch with hands against the wall, one leg bent and the other to behind it to be stretched. She said hold for 45 seconds. Do 3 reps. Do it 3 times per day. The 2nd is balance. Simply stand on one foot (ya, right, LOL). The goal is to hold one foot up for 10 seconds. If needed, place fingers on a table or chair back as needed and wean off over time (if possible). Do 3 reps. Do it 3 times a day. She said do this barefoot, so it also

stretches the Achilles and the hip, which is good. The 3rd is to stretch the back of the leg. Sit, with one leg stretched out straight ahead, with heel on top of a foot rest. "Dig" the heel into the foot rest, so that the back of the thigh and calf are taut. Hold 45 seconds. Do 3 reps. 3 times. I asked her about standing and doing toe touches as an alternative, as she said that would be good, too.

**AAC-RERC Augmentative and**

**Alternative Communication Technologies - Rehabilitation Engineering Research Centers**

<http://www.aac-rerc.com/aac-rerc.htm>

Mission & Objectives The AAC-RERC conducts a comprehensive program of research, development, training, and dissemination activities that address the NIDRR priorities and seek to improve technologies for individuals who rely on augmentative and alternative communication (AAC) technologies. The mission of the AAC-RERC is to assist people who rely on augmentative and alternative communication to achieve their goals by advancing and promoting AAC technologies and supporting

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services to rural and urban areas, and (2) other scientific research to assist in meeting the employment and independent living needs of individuals with severe disabilities; or Facilitating service delivery systems change through (1) the development, evaluation, and dissemination of consumer-responsive and individual and family-centered innovative models for the delivery to both rural and urban areas of innovative cost-effective rehabilitation technology services, and (2) other scientific research to assist in meeting the employment and independent needs of individuals with severe disabilities. Each RERC

the individuals who use, manufacture, and recommend them. What is an RERC? Rehabilitation Engineering Research Centers (RERCs) carry out research or demonstration activities by: Developing and disseminating innovative methods of applying advanced technology, scientific achievement, and psychological and social knowledge to (1) solve rehabilitation problems and remove environmental barriers, and (2) study new or emerging technologies, products, or environments; Demonstrating and disseminating (1) innovative models for the delivery of cost-effective rehabilitation technology

must provide training opportunities to individuals, including individuals with disabilities, to become researchers of rehabilitation technology and practitioners of rehabilitation technology in conjunction with institutions of higher education and nonprofit organizations. The RERCs work at the individual level focusing on technology to lessen the effects of sensory loss, mobility impairment, chronic pain, and communications difficulties. They also work at the systems level in such areas as eliminating barriers to fully accessible transportation, communications, and housing. Partnering with industry, product

developers, private sector entrepreneurs and even hobbyists, the RERCs embody the potential to make sweeping changes affecting public policy and the nature of the built and virtual environments. For more information about RERCs, visit the [National Center for the Dissemination of Disability Research \(NCDDR\)](#) (800) 266-1832 or (512) 476-6861 National Center for the Dissemination of Disability Research Southwest Educational Development Laboratory 211 East Seventh Street, Suite 448 Austin, Texas 78701-3253

### **Finding Acceptance of Ourselves and our Disabilities**

Rev. Nancy Lane, Ph.D Know who you are. Accept who you are, without shame, without apology. Integrate your limitations and disabilities into the whole of who you are. We are all different, all of us unique. Do not blame disability for everything that goes wrong in your life. Everyone struggles with loss, problems, suffering, and eventually death. At the same time, recognize that a struggle for acceptance and value in society is involved. We are participating in a universal struggle for acceptance of the diversity which is found in the image of God. The responsibility for yourself, your life, and your growth is yours alone, and

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trusting our inner responsibility to being who we are, as we are.  
Acceptance is found only by letting

no one else's. The "blaming game" is a dead end, vicious circle that leads no where except to more of the same. We heal the wounds as we name and acknowledge their affect on our lives; forgive ourselves, others and God as necessary; and let go of the pain. Allow yourself to enter into the moments of anguish, depression, failure, and grief. We do experience loss; we do grieve for real or perceived images of who we were or might have been. The grief cycle will recur throughout your life time. One of the most debilitating neuroses of our time has been defined as being the desire to be perfect. This is the trap of the "tyranny of the should's," which says: We should be honest, courageous, brave, unselfish. We should be able to endure everything, be like everybody, love everyone; nothing should matter; we should never feel hurt, or feel sorry for oneself; we should be grateful to be alive at all times; we should be in control of all emotions and feelings; we should not wish for things to be different; we should be able to overcome every difficulty. The "shoulds" are a pressure against us determined by a world alien to whom we are. As long as we "should," we are serving the gods of others and not

go of our "idealized self-image" which denies our limitations and problems. Acceptance is letting go of

all guilt and shame. People who abuse, discriminate, and are otherwise insensitive, unaware, or inaccessible do not apologize for their behavior. Acceptance is not "out there" somewhere: it is to be found deep within ourselves. If we cannot accept who we are, we cannot expect others to do so. Focus on the abilities you have been given, and not on what you have lost or never had. Acceptance is about living with disability in creative and meaningful ways which celebrate the goodness of who we are, claiming your gifts and abilities and then using them creatively to make a difference in the world around you. Simply accept the fact that you are accepted.

**Speech Loss Survey** Requested by Linda Gentner The ALS Clinic at UCSF wants answers to some speech questions: What was your voice like before you lost the ability to speak?

(1) soft - slow - slurred (2) strained - coarse (3) or what?

Please reply to Linda Gentner

[lkgentner@aol.com](mailto:lkgentner@aol.com) or

510-651-5676

**When It's Time to Take away the Keys** Contributed by Lissa Robins Kapust, a licensed clinical social worker at Beth Israel Deaconess Medical Center, Boston. *Ed. Note - the following is excerpted from a*

*letter to the editor which appeared in the Boston Globe in response to a series the paper ran on safe driving.*

Americans have complicated love affairs with their automobiles and many feel that you are what you drive. When the question is raised about whether it is

time to give up the keys, this triggers a crisis for individuals and families. In my role as the program coordinator for a hospital-based program that focuses on the driving competence of individuals with medical and/or age-related problems, I understand the tugs and pulls around resolving a driving issue critical to independence and emotional well-being. We need to remember that driving is not a right; rather, it is a privilege -- one that should be afforded only to those who can demonstrate responsible and safe driving behaviors.

**Estate Concordia on St. John for Disabled Travers** By Claudia

Dreifus When Stanley Selengut started losing his vision to macular degeneration three years ago, he began thinking about how his ecologically oriented resorts in the U.S. Virgin Islands could better accommodate people with disabilities. If someone's in a wheel-chair," says Selengut, an ecotourism pioneer who's now 76 and legally blind, "most of what's available to them now are cruise

ships and a few designated rooms at mass- tourism hotels." Working with Multi, Design for people, a Rhode Island-based studio, Selengut revamped plans for his newest venture, Estate Concordia on St John. In five wood-and- canvas "ecotents" he had special ramps and railings installed, as well as extra-wide bathrooms, open shower stalls and easy-to-use fixtures and appliances. His design team also worked with local merchants on sporting and entertainment venues.

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kind of vacation," reported one of them, Ileana Rodriguez, 19, a student from Miami who's paraplegic. "What Mr. Selengut has done here will hopefully be a model to other resort operators." Contact: Kat Darula or Melody Smith Multi, Design for People, LLC Maho Bay Camps, Inc

[kat@designforpeople.org](mailto:kat@designforpeople.org)

[melody@maho.org](mailto:melody@maho.org) T: 401.935.2976 T: 340.693.5722 x233.

**Living Alone** *Ed. Note: I said the following in a posting when my husband Jim was away," . . . I don't know how single parents raising kids can do it, or those of you living alone and working full time. You have my deepest respect and admiration." Here are two replies I received from PLSers who live alone. "Your posting about being alone was very*

Taxi drivers were given portable ramps to make their cabs wheelchair- accessible. Plastic mesh mats were set down on beaches, making it possible for wheelchairs to reach the water. And thanks to a lifting device called "Love Handles" tourists can now board boats for snorkeling, sailing and kayaking. In July, Selengut invited four travelers with disabilities to come to Concordia to test everything. "I never thought I'd have this

validating. I live alone with Frick and Frack, my two littermate tuxedo cats, and realized that when G-d was giving out attributes....I must have skipped the line of "patience." It takes me so long to get myself ready in the morning that it is time for lunch and I haven't eaten breakfast. I have an aide 5 x/week from 4-7 PM. They take me to doctor's appointments, unpack my curb sided pickup of groceries, and make dinners, lunches, breakfasts and put them in the freezer. Then I can just pop them in the microwave. It takes me so long to do things that sometimes I decide that it is not worth it. I find the weekends the most difficult because I can drive to places but not walk from the car to my destination. My PT, my hairdresser and some doctors will send out someone with a wheelchair for me....but otherwise it

is really so ludicrous that I can drive (and safely) but not walk. I'm beginning to lose sensation in the tips of my fingers which has caused many a dropped thing....last week a plastic container of walnuts. It took me 45 minutes to clean it up because I didn't want the cats to play soccer with the walnuts. M. Scott Peck in his "Road Less Traveled" refers to people being "Human Doings" or "Human Beings" and I've always felt validated by my "doings". It is so hard at 69 10/12 years old to shift into another gear. I try very hard to focus on the half of a cup that is full...not always easy." Bebe

"I have rarely said anything when the subject of living alone comes up because I don't know how to get to the nitty gritty of the subject with neither complaining nor bragging. I just know it's not easy being alone and from feedback I get at work and family reunions from folks I only see a couple of times a year I must be making it look a whole lot easier than it is. I'm glad I don't have some of the added problems that others do, but not being able to hear certainly

makes life alone an adventure. Last week when we were expecting Daddy's death it was terrible. I don't think my own intimate family gave it a thought the way I did. I couldn't stay at Mother's because I can only sleep in a recliner and it is too much trouble to drag the TDD around with only one hand so I would have no phone access. If I were home alone nothing wakes me once I remove the hearing aides. I was power of attorney so I needed to be accessible. I left my house open a couple of nights because I didn't want my extra keys to get scattered and I sure didn't want my garage door code floating around heaven knows where. It would have gotten passed along and I never would have known who might just walk in. Some of these people DO NOT THINK. I just said it wasn't working....My niece came home with me but I know she wanted to be with her grandmother. I've said it before and I'll say it again---It ain't easy being me....or you or any one of the rest of this bunch." Ronnie

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toilets were the normal sort so I soon learned that I had to hold on until I saw a sign for a disabled toilet - or else headed for western style hotels

whenever I could. We've come across many challenges with different power outlets. One new hotel in China had power points that

could take all sort of different shaped plugs - now that's what I call sensible. Why can't hotels do that everywhere?

Contributed by Gary Duke My Dad offered us his time share week in Panama City Beach-Florida along with cash (because he knows we are on a budget). So my wife and I drove down there on the 10th and had a wonderfully warm. The funny part is that it was College Spring Break. Our condo was more elderly and families, but next to ours was a Holiday Inn. They had a stage set up on the beach (for their bikini rub downs and dancing and such). They had a mechanical bull set up and lots of beer. Out side the "spring break world" there was a radio station that played rock-roll and offered up prizes for games they would play. MTV was there and they even had paratroopers from Fort Bragg drop in. It was so funny watching these kids get drunk and act stupid. Lots and lots of girls in bikinis. My wife said that if I was to die next year I probably would still have a smile on my face. There were so many drunks staggering on the beach. And then there was me with PLS and I couldn't even walk a straight line from the deck to our chair rental. I know everyone thought I was drunk. Then to top it off we went walking Wed. night on the beach. My legs were feeling strong so I said to my wife,

"I'm gonna run and see how it feels". She of course tried to discourage me, but I ran anyway. Yep-I took 4 steps and was eating dirt. I fell face first and was laughing so hard at my stupidity and then I couldn't even push myself up. As I went staggering away I could hear people laughing at me

**Travel Stories** Contributed by Kiwi Linda Our youngest son got engaged last year to a fellow student, studying here in New Zealand. She came from China. So in December we all went to China for a traditional Chinese wedding. It was wonderful. I got to walk on the Great Wall. This was amazing. I felt so grateful to be able to do it. The place where we went had a cable car where you got most of the way up -but then you had to walk on some uneven ground and up some very steep steps. I would never have made it on my own - but with a helping hand I managed to get onto the wall and walk one section. Getting down was quite a challenge - this took two helpers and I had to go down backwards. So I backed down the steps with my husband below me and a son above to help. Must have looked quite a sight but I DID IT. Traveling on planes in China was sometimes a challenge as sometimes we had to go down steps to the ground - and the last step can be a long way away from the ground. This left me dangling a couple of

times and I had to get help and then there might be a high step up into a bus - so lots of times I needed a heave up. So not to be recommended on your own. I did use a cane some of the time in China. Then it was onto the wedding in Shenzhen. The young people could speak some English as they learn it in school. I was really proud of my family - everyone looked great and mixed well with the people we met. We ate lots of different foods. I

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on the balcony. I know what they were thinking - "Stupid drunk"!

Contributed by Linda Gentner We recently returned from the British Virgin Islands (along with Thurza and Jim Campbell and another couple) and Thurza and I both fell on the beach coming out of the water. Thurza and Jim were in the water by themselves so she said Jim had to drag her across the sand and, of course, being the way we are, she laughed, and became helpless. I went into the water after much coaxing and, yep, coming out--even with Craig and Jim on each side--down I went and of course I couldn't stop laughing but I guess that was better than crying. Loved the warm weather so it was worth the fall!

Contributed by Thurza Campbell  
More on the vacation of two PLSer

have trouble eating with a knife and fork so just couldn't manage chop sticks, so if there was no fork I ate with my fingers. I had a wonderful Chinese massage and I came home feeling really great. I wouldn't have missed it for the world. I'm so grateful I was still able to do it. The places we went wouldn't have been accessible in a wheelchair. Most of the public toilets in airports and such are the squatting sort. Fortunately "disabled"

couples. The slogan for the week was, "hey mahn, go slow, be happy, no worry." We had VIP treatment at Logan airport in Boston from curbside to plane, which was great, with me in a wheelchair. At the resort, Craig lovingly takes breakfast to Linda in their room before even having coffee! I am impressed! We took a catamaran ride to the Baths at Virgin Gorda. Everyone but Linda and Thurza went topside to hear the stories of the Baths. One might wonder why the lecture couldn't have moved downstairs so we could have heard it, and were we aggressive- handicapped types, we might have gently asked...but maybe someday someone will tell us the stories. Snorkeling for Thurza back on Tortola had a couple of traumas, due to gear adjustments, inability to communicate since I cannot speak, my sheer panic, and being pushed forward while swallowing the entire

Caribbean Sea, but I survived, and am ready to try again. The underwater sights are unforgettable. On St. John we went to Mongoose Junction. There are steps everywhere.

Oh, for the days when steps weren't an issue!! For Linda and Thurza, thanks to the able assistance of our Human Durable Medical Goods, aka Jim and Craig, we take turns having them on each side. We can go anywhere, do anything, conquer the world! Back at the San Juan airport a motorized cart took 4 of us to customs. San Juan airport is pretty well adapted for handicapped, even though Linda and Thurza did go up two escalators. The only mishap flying home was Thurza's walker. . . Dx broken leg. "hey mahn, go slow, be happy, no worry." American Airlines reimbursed us for a replacement. Flying out of the US does work for handicapped; be sure to pack both your sense of adventure, and your sense of humor.

**As Luck Would Have It** By Michael Shermer  
Contributed by Alper Kaya

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Hawking, who writes: "I was lucky to have chosen to work in theoretical physics, because that was one of the few areas in which my condition would not be a serious handicap." Unable to move and confined to a wheelchair, Hawking has capitalized

MD Victims of ALS could be forgiven for feeling unlucky. How, then, can we explain the attitude of the disease's namesake, baseball great Lou Gehrig? He told a sellout crowd at Yankee Stadium: "For the past two weeks you have been reading about the bad break I got. Today I consider myself the luckiest man on the face of this earth." Clearly, luck is a state of mind. Is it more than that? Richard Wiseman created a "luck lab" at the University of Hertfordshire in England. He found that though lucky people were twice as confident as the unlucky ones that they would win the lottery, there was no difference in winnings. He further found that a satisfied state of mind can translate into actual life outcomes that someone might call lucky. Expectation also plays a role in luck. Lucky people expect good things to happen, and when they do they embrace them. But even in the face of adversity, lucky people turn bad breaks into good fortune. Consider the example set by one of the longest ALS sufferers in history, Stephen W.

on his fate by using it as a chance to transform our understanding of the universe. *These interesting clips are some of the comments Alper's post generated.* . . . My "twin" continues to teach me new paradigms. He was born with cerebral palsy. He is

brilliant and is months away from retiring as a civil engineer and highway designer for the state of West Virginia. He has a weak left side but walks with a limp. He drives an Audi TT two-seater convertible with a standard shift and jokes about what people say as they walk past his car in the handicap space. His theory is positive and loving thoughts attract positive and loving karma which attracts all that is positive and loving. I haven't mastered the whole package or I wouldn't be whining to this group as I have post surgery. However, I did make it to Florida solo and have done some pretty amazing stuff mixed in with all the 'bad luck.' Lol. If you are able to be happy in your mind above the pain what else really matters. Life IS an Attitude!" Eva

"I think that using the term "luck" implies to me that there is no control over our destinies and our daily dealings with what life throws our way. There have always been major stressors to overcome in my life and PLS has become just another one of those things. It is interesting the researchers chose to use "luck" as their paradigm. We all have challenges to some degree or another. In my experience, it is gratitude that pulls me from asunder the darkness. I sometimes need to see this thru friend's eyes first, but am getting better at seeing it in my

life all by myself (I am a grown up now). That said, PLS has given me the greatest gift and blessing of all of the shit before it. PLS has allowed me to be still. In my life prior to getting ill I was working on being still and living in the moment and trying to leave work at work and not work so much, etc., and although the strides were small, I was proud of them. Then came this yucky, dark, sudden, rapid PLS monster that I felt certain would be the thing that would send me over the edge, when this functioning left and then that and then that and still again that.....each time tests my ability to transcend the experience so that I may enjoy this life as much as I can. Do I consider myself lucky? I cannot say that I do. I consider myself stronger and more determined to live this life rather than just surviving it. I am actually grateful for the ability to be still and figure out I really can paint, I can appreciate something moving slower than the speed of sound, I am ok without saving people at work. Of course that could be construed into lucky, if I only knew what the definition of luck was!! It is up to me to plow my course from here and something tells me that luck has nothing to do with it!" Tawny Swain Castle

**The Journey** by Karen Rossman (DFCI Newsletter) There are incidents in all our lives that change

our paths forever. The birth of a child, death of a loved one, a devastating diagnosis. In these moments, time freezes for us, and it is incomprehensible that the outside world continues on as if nothing has changed. They alter the blueprint on

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some of which we are grateful to have had, all of which influence us on our journey through life. But it is our reactions to these events that make us who we are; how we handle the circumstances we are dealt. It is often through the smoke of the aftermath that our true purpose is unveiled and our most important work done. As we struggle through, we are often inspired to create the best we have to offer, as our true gifts are unleashed from within us. It is only at the journey's end that we can look back and see how far we have come on the path that was our destiny to have traveled. Will we make the choice to improve the road or stop in our tracks and refuse to grow?

**My Ideal Perfect Day** *While watching "Tuesdays with Morrie", Mitch posed the question to him, "what would you do if for one day you were healthy?" The idea haunted me, as it made me realize what I miss the most. Writing it down helped me focus, then move beyond the losses. Thurza* Instant thoughts burst into my consciousness -

our souls, and the roadmap for our lives. They challenge us to grow and develop in ways we never dreamed. They bring out our good, our bad, our hope and our fears. They contain our life lessons, many of which we never would have chosen,

To ski top speed down Avalanche in powder,  
perfect turns, feel the invigorating sting of the cold wind in my face, unweighting like flying. And I'd talk, talk, talk. But if it's summer, I'd body surf.  
Beat Jim and the kids, of course. And I'd talk, talk, talk. Then steak  
a thick, rare one sink my teeth blissfully into the firm texture and chew. And I'd talk, talk, talk. Later, I fell asleep planning my entire perfect day -  
not just knee jerk instant flashes,  
but careful reminiscing. I'd toast and revel in eating a crusty, chewy bagel  
And I'd talk, talk, talk. I'd spend the morning transplanting shrubs  
And weeding - really! All afternoon I'd sing at nursing homes with the Music Makers. Then end with a wonderful party here -  
I'd prepare all of the delicate fancy food.

I'd circulate among our guests with trays.

And I'd talk, talk, talk. I'd laugh and engage in conversations with family and friends

Keeping up Sometimes sparring Being clever Being a perfect hostess. Why'd I write this? Such simple, everyday things I'd do in my perfect day.

A reader'll think me nuts. But--- Might this help you consider that the simple things

You take for granted Are blessings to treasure.

Carpe' diem!

## CAREGIVING

### New Appreciation for Caregivers

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my waking hours!! I don't know how single parents raising kids can do it, or those of you living alone and working full time. You have my deepest respect and admiration. And to Jim, now that I realize how much you do, another "thank you". Thurza

"It's easy to not notice how much others do as their part of the team when their part increases gradually. And I, too, am in awe of people with our challenges who live alone. How do you do it? And that's a real question I hope people who live alone will answer. I need to get more comfortable being alone as it looks

"Since my dx in '01, and my steady progression, Jim's gradually helped me more and more. Especially he's had to do more in the last year as my hands have become affected by PLS as well as my voice and legs. Well folks, I had no clue just how slow I'd gotten at ADL (activities of daily living) until I started doing them solo when Jim was on a trip. Since Jim's increased help had been gradual, I didn't realize how significant it had become! Wow! I have two emotions now - gratitude for all he does, and awe for those of you who live alone!! It's incredible that ADL can take up most of

like Ed will be traveling more with his added on responsibilities." Kathi

"Craig's father is having surgery in a couple of weeks so he'll be gone for a few days and then he'll be back for one day and then he'll be gone on another trip for two more nights. I have Molly & Sadie (dogs) to keep me company but no family real close. We have friends who always offer their help but I try to appear to be more independent than I really am. So...I'm not looking forward to it but, he is needed elsewhere. I realize all he does for me. What I find interesting is that the dogs don't

expect me to walk them but they sure do Craig. I too, have always admired the people who need to live alone with a disability. I hold them on a pedestal." Linda

"When I was DX back in 1986, the doctor said "a form of Lou Gherig's Disease", we were devastated and scared. There was no PLS Friends and I felt so alone and scared. Did have a clue what was down the line for us, so we sold our home of 25 years and bought a modular home in an adult community park. Everything was on one floor and in the beginning I could take care of it along with my darling husband. But as my conditioned worsened it was John

who decided I had to be in a place like this, as it was getting more difficult for me, and my balance was really out of whack. We found this place and as much as I hated to give up my home we are here now, and it's where I (not we) belong. This is when I met my first PLS person, Mark Weber and the rest is history. Yes Thurza I am also a lucky lady (as you know) I can't imagine not having him here to do what he does for me 24/7. Does Jim pluck your eye brows?? John does (he would kill me if he knew I told the world this) LOL! The reason we love living here in our senior citizen community is that if anything were to happen to

him, I would have 23 other additional caregiver's, as that just what we do here, nobody is ever alone.

Sometimes I say to him, why do you stay with a person like me, and he get soooo mad at me, but its how I feel. He should be enjoying these years instead of catering to somebody in my condition. I think sometimes we forget about what they go thru watching us slide down to what and where we are. Oh well enough of that!!!!" Rita

"Yes we guys can be good, but it is the ladies that are caregivers that sometimes take the worst. It is hard on the men /women that are still working and caring for someone. John and I are retired and it gives us something to do. Carrie has hand problems. I get her eyebrows. I trim her nails and toe nails. She polishes the nails with an electric buffer and polishes one hand with polish and I have to do the other and the second coat on both. When she wants a bath that is also my job and we have our little routine. She can manage to transfer to a chair in a special shower and back to stool get dry and a nighty. I cut her hair when needed and do better that most salons do. I use a flowbee which is a cutter with different heights and hooks to a tank vacuum. The family says I do a better job, but she

won't admit it LOL. The only thing I will not do is try to get her off the floor. She slid out of her scooter in the bath room and banged up her foot real bad. I just called 911 and told them no sirens or lights and there were about 7 men on 2 big trucks and two picked her up and put her back in chair and examined her foot and no broken bones. I thanked them and they said we do it quite often. Good luck to all and I think more people should talk about problems and what has to be done for the other. Carrie would be like John if she knew all this was on e-mail. But it may help others."  
Carrie and Byron in Sanford

"I consider myself one of the lucky ones too. Although I can still do most things by myself, there is still plenty I can't do, like hang out the washing (aussies mostly all have clothes lines in their backyards...its an aussie thing) take out the garbage, do the big grocery shopping, mop the floor, sometimes vacuums, lift heavy stuff, cook when I am tired etc etc. I thank the Lord that I have a wonderful husband who does all that and more. He nearly died 3 years ago and was sick for 2 years after that but he still kept doing all his chores. I have often wondered how I would cope without him. I know that I would learn to cope by getting outside help but its much easier and a comfort to

have him by my side. He has a great sense of humor and makes me laugh every day. I tell him always that he has to outlive me. But having said all these wonderful things about him, I don't know how HE would cope if the going got tougher with me with personal care etc. I don't think about that hardly at all because I believe I will stay the way I am for a long time. Here's hoping! So I take my hat off to all of you here that are coping on your own. And the same goes for the caregivers too!!!" Aussie Maureen

"I tell my husband he can not die before me that I have to go first as I can not do anything without him. I think he does worry about this."  
Jane Anne

**A Tribute** Kathi Geisler In December of 2003, a man with PLS named Gerry Leary and his wife Barbara attended a Lunch Connection that I organized in MA. There were a couple of dozen of us funny walkers there, with either HSP or PLS and lots of spouses. Gerry was still working as a financial advisor and had recently begun to use a cane. He said he started to have walking problems many years ago. He was a charming man with a great smile and vivacious personality. His wife was also charming. Since then, they had attended the various events we've had in and served on the National Conference/TeamWalk Committee.

When our group met for an outing just over a year ago, two years after I had met him, you could really see how his condition had drastically changed. Barbara was retiring from work early, so she could have time with him. They moved into a condo in a new over- 55 development with the master bedroom on the main floor. Despite everything, Gerry maintained the same twinkle in his eye. Last winter ('04) Barbara planned a trip to Key West for 6 weeks. There was an extraordinary amount of planning and organizing

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day. Every time he needed to use the bathroom, she transferred him from the lift chair to the w/c to take him in. He could still speak a bit, but it was belabored. Despite it all, he still had the twinkle in his eye and smile and we had a lovely time there. It was clear that these were not easy days for them, but that they were both doing everything they could to make themselves as comfortable as possible. Back home, health services were set up to come each day to give some help.

Barbara's job was still 24-7 and it became clear that the time had come for the hard decision to move him into a Hospice. Gerry passed away in February. He was 62. I spoke at the reception about Gerry's involvement in our group and what a remarkable man he was and what a

Barbara had to do for this trip because of Gerry's serious needs. She had to rent a lift chair for him and rent or buy lots of special daily living aids, mobility devices and bathroom items. She set up medical services so specialists could come to the condo. We visited them for dinner. Barbara had to transfer Gerry from his bed to a roller seat to take him in the bathroom to get him ready for the day. From there he went into the w/c and from the w/c to the lift chair in the family room each

champion he was with his illness and also, what a remarkable wife he had. I never would have met Gerry if I hadn't had HSP and he hadn't had PLS. I don't wish these conditions on anyone and hate that I have it, but there has been a silver lining....I have met remarkable people like Gerry and Barbara whom I never would have ever met. I have found out how wonderful people are and how important we all are to one another.

## **MEDICAL RESEARCH**

**What's Happening in PLS at Northwestern.** Nailah Siddique RN MSN Neuromuscular Disorders Program, Northwestern University  
The most exciting thing is that we

are in the process of hiring a nurse just for PLS! It has taken us a very long time to find the right person, but finally we have. She has been running another research project within Northwestern in preventive medicine, so she has the right kind of experience and she is very interested in PLS. There are a few hoops to be jumped through before she can actually begin, but we expect there will be a nurse devoted solely to PLS available to you by mid-April!! In the meantime, this is what she will inherit. We have been collecting blood samples from PLS patients and appropriate family members for several years now and have the nucleus of a good sample set with 170 patients and various relatives. Additionally, we are starting collection of CSF from patients for proteomic studies. We currently have CSF samples from 15 patients, 4 of whom have PLS. We have 3 more PLS samples pending planned for the next month. We have elected not to obtain samples from 3 people who offered who are using a Baclofen pump. The scientist doing the proteomic studies feels we need a little more baseline information

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two other papers from the meeting published in the journal "Amyotrophic Lateral Sclerosis and other Motor Neuron Disorders", one by Rowland, the other by Strong and Gordon. And, as hoped, the meeting

before we add drug and drug byproducts to an already uncertain and complex picture. Thurza asked me to detail how we go about getting the CSF samples. A patient offers to have an LP done, or share CSF when (s)he is already going to have a tap. I discuss the plan with the MD who will be doing the tap to see if the MD is willing and arrange payment to the MD if necessary. Our consent form, tubes and shipping materials are sent either to the patient or the facility doing the tap. Then the sample is sent here on dry ice. We ask for 10 mls of fluid, but accept anything that can be donated, so sometimes our samples are as small as 2 mls. Unfortunately, we don't have the funds to pay people for donating their CSF. We use the money we have to make sure it doesn't COST a person anything to do it! Generally, the donations have gone smoothly. There are several reports of the PLS meeting held in June, 2004 in California. This is the most complete summary available. It was published in "Neurology Today", a publication of the American Academy of Neurology. November, 2004. There have also been

also engendered follow-up work, including the recent exciting paper from the group at Columbia published this month in "Neurology". Lastly, I'd like to say thank you to all of you who have borne with us while

we're getting started with the Registry. I think it will move forward much more rapidly with our new nurse.

**MDA Research Notes ALS Connection** is a patient driven registry to collect data from ALS patients in North America. The project receives funding from MDA and is being coordinated by Drs. Robert Miller, Jonathan Katz and Catherine Madison, all physicians who treat ALS patients in a multidisciplinary clinic (The Forbes Norris MDA/ALS Research Center) in San Francisco, CA Programming staff of the Department of Epidemiology, University of California, San Francisco (UCSF) developed the database and website and monitors them to ensure complete confidentiality of the data. ALS Connection was created to: - identify opportunities to improve the quality of care for patients with ALS; - learn more about the disease evolution; - obtain long-term follow up data and information about quality of life as well as outcome of patients with ALS - provide patients, caregivers and healthcare providers with information about ALS and ALS related topics - provide data for ALS research. Anyone who has been diagnosed with ALS by a physician can enroll into this registry from the website [www.alsconnection.com](http://www.alsconnection.com) . The information that the individual

provides will be used to evaluate variations in patient care and adherence to standards of care, as well as help foster ALS research. An additional focus of this website is to educate participants and visitors to this site about ongoing ALS research. Other features of the website include: - information about studies and an explanation of what a clinical trial is - meeting summaries; - educational resources; - additional web links for health care professionals.

**MDA Launches ALS Translational Research Program** MDA has launched a new translational research program specifically for [ALS](#). The term "translational" describes moving strategies that are ready to leave the laboratory through the appropriate administrative and regulatory pathways into clinical trials. Translating research from "bench to bedside" can be difficult, because scientists aren't always fully aware of how to move their cell and animal studies into the clinical arena, and physicians aren't always fully aware of what's going on in academic laboratories. MDA's general translational research program, which eases the way for such transitions, has been highly successful in moving gene re-reading strategies for Duchenne

muscular dystrophy and myostatin protein blocking strategies for adult muscular dystrophies out of the lab and into testing in patients.

### **Copaxone Results Encourage**

**Investigators** Paul Gordon, associate medical director at the Eleanor and Lou Gehrig MDA/ALS Center at Columbia University in

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six-month trial of Copaxone, a drug approved for use in multiple sclerosis. Gordon, who hopes to take trials of Copaxone in ALS into the next phase, says the results are meaningful "because we showed that we can alter the immune system in ALS patients the same way that it's done in patients with multiple sclerosis, where the treatment is effective in slowing the course of the illness. Whether these changes will correlate with clinical outcomes needs to be determined in trials powered to assess clinical efficacy."

### **MDA Supports Bill to Ease**

**Biotech Restrictions** MDA has joined some 30 other organizations that fund medical research in urging Congress to ease restrictions on small business grants for new biotechnology companies. Recent changes in the government's Small Business Administration grants program have excluded companies that rely heavily on outside investors

New York, and colleagues, have found that *glatiramer acetate*, better known as *Copaxone*, was safe and well tolerated in people with ALS, and that the participants showed evidence of altered immune-system activity with the drug. 30 people with ALS participated in this

(venture capitalists), which describes most biotech startup firms. The Biotechnology Industry Organization (BIO) is sponsoring a bill (H.R. 2943 and S.1263) to lift this restriction.

### **Helpful Terminology Gene**

-The basic unit of heredity composed of DNA that is the code for directing the production of proteins. Genes exist in pairs, one on each of the pairs of chromosomes. **Gene**

**Therapy** - An experimental process that seeks to replace a defective gene to correct an inherited disorder. When the procedure is performed on cells outside of the body, it is known as "ex vivo" gene therapy.

**Genome** -The term used to refer to all of the genes found in any individual. **Neuropathies**- affect the nerve cells in the brain or spinal cord, or the nerves that run between the brain and spinal cord and the rest of the body.

**Myasthenias**-affect the place where nerves meet muscles.

**Myopathies** -affect muscles.

**Antisense Therapy for ALS** A pilot study will test a new therapeutic approach called antisense now being applied to ALS. ALSA funded the basic research that demonstrated the ability of antisense molecules to improve ALS symptoms in rats modeling the disease, noted Lucie Bruijn, Ph.D., ALSA science director and vice president. The antisense pilot clinical trial will be guided by principle investigators Merit Cudkowicz, M.D., Massachusetts General Hospital in Boston and Richard Smith, M.D. of the Center for Neurologic Study in La Jolla, Calif. Timothy Miller, M.D., University of California, San Diego is a co-investigator with Dr. Smith and was instrumental in the preclinical work that has led to this trial. Antisense targets the message to produce a particular protein and acts directly on RNA, the message molecules that spell out the instructions from the DNA of the cell's genes, in essence

Synapse - Spring 2006 Edition Page 19 established in this study, and efficacy shown by subsequent studies, the stage is set for development of additional antisense-based drugs for the treatment of sporadic ALS, provided the appropriate molecular targets are identified.

**Research snippets Scientists**

interrupting the message. (First generation antisense agents are now further improved to dramatically increase their persistence in the body and their therapeutic effect. More than 500 people have received these second generation antisense candidate therapies in clinical testing in a variety of diseases, suggesting it will be safe for ALS patients, but such safety must be shown by this pilot clinical trial). If successful, antisense technology is at a stage of maturity that permits the development of additional compounds targeted to different proteins identified as important in a disease process. These products can readily be manufactured at market scale in FDA-approved manufacturing facilities, the investigators noted, and cost of these therapies will likely be similar to or less than current biological products such as interferon. Should tolerability be

**Crack Motor Neuron Code** Cell, November.4, 2006 Columbia neuroscientists have deciphered a motor neuron code that one day may help in attempts to restore movement in people with diseases like ALS and Spinal Muscular Atrophy (SMA) or spinal cord injuries. Motor neurons within the spinal cord in patients with ALS and

SMA die over time. Thomas Jessell, Ph.D., says "We will have to understand how to connect the neurons to the right muscles in order to restore movement." The connections motor neurons make are established during development. This wiring process is not random -each motor neuron has already formed an allegiance to a particular muscle before the axons start growing. "The more we understand the basic workings of the entire locomotor circuit, the better chance there is of developing regenerative strategies to restore movement." The research was supported by the NINDS and Project ALS.

**ALS and PLS: The Role of Diffusion Tensor Imaging and Other Advanced MR-Based**

**Techniques as Objective Upper Motor Neuron Markers.** Wang S, Melhem ER. Department of Radiology, Division of Neuroradiology, Hospital of the University of Pennsylvania, 3400 Spruce Street, Second Floor Dulles, Philadelphia, PA 19104.

[emelhem@rad.upenn.edu](mailto:emelhem@rad.upenn.edu).

SOURCE: Ann N Y Acad Sci. 2005 Dec;1064:61-77.

<http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list>

[\\_uids=16394148&dopt=Abstract](#)  
Amyotrophic lateral sclerosis (ALS),

also called Lou Gehrig's disease, is a motor neuron disease characterized by progressive degeneration of upper motor neuron (UMN) and lower motor neuron (LMN), while primary lateral sclerosis (PLS) is defined by pure UMN involvement. A reliable objective marker of UMN involvement is critical for the early diagnosis and monitoring of disease progression in patients with ALS and PLS. Diffusion tensor imaging (DTI), magnetization transfer imaging (MTI), and magnetic resonance spectroscopy (MRS), which provide insight into the pathophysiological process of ALS and PLS, show great promise in this regard. Further investigation is needed to determine and to compare the utility of various neuroimaging markers.

**Rapid Progression of Scoliosis Following Insertion of Intrathecal Baclofen Pump.**

Sansone JM, Mann D, Noonan K, McLeish D, Ward M, Iskandar BJ. From \*Department of Neurological Surgery, University of Wisconsin, Madison, WI; and dagger Department of Orthopedics and Rehabilitation, University of Wisconsin, Madison, WI. J Pediatr Orthop. 2006 January/February;26(1):125-128. Placement of an intrathecal baclofen pump is a common treatment of spasticity in cerebral palsy patients. Though effective, the hardware is

prone to malfunction, and multiple revisions are often necessary. Additional complications have also been described, including infection and drug toxicity or withdrawal. The authors report another complication of intrathecal baclofen therapy:

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with any patient considering  
intrathecal baclofen pump

**Botulinum toxin treatment of adult spasticity : a benefit-risk assessment.** Sheean G. University of California, San Diego, California, USA. SOURCE: Drug Saf. 2006;29(1):31-48.

<http://tinyurl.com/bkydq> Injections of botulinum toxin have revolutionised the treatment of focal spasticity. Botulinum toxin can produce focal, controllable muscle weakness of predictable duration, without sensory adverse effects. Randomised clinical trials (RCTs) involving patients with spasticity resulting from a variety of diseases have clearly shown that botulinum toxin type A (Dysport((R)) and Botox((R))) can temporarily (for approximately 3 months) reduce spastic hypertonia in the elbow, wrist and finger flexors of the upper limbs, and the hip adductors and ankle plantarflexors in the lower limbs. There are very little data to allow a benefit-risk comparison of phenol and botulinum toxin injections; each have their clinical

accelerated progression of scoliosis after pump insertion. Although there may not be a direct correlation between intrathecal baclofen and scoliosis, the authors consider the issue important enough to warrant discussion

and technical advantages and disadvantages, and phenol is much less costly than botulinum toxin.

**Hereditary Spastic Paraplegia** Dr. John Fink Department of Neurology, University of Michigan and the Geriatric Research Education and Clinical Care Center of the Ann Arbor Veterans Affairs Medical Center, Ann Arbor, MI 48109-0940, USA. [jkfink@umich.edu](mailto:jkfink@umich.edu) ; Curr Neurol Neurosci Rep. 2006 Jan;6(1):65-76.

<http://tinyurl.com/8pe79> The hereditary spastic paraplegias (HSPs) comprise a large group of inherited neurologic disorders. HSP is classified according to the mode of inheritance, the HSP locus when known,

and whether the spastic paraplegia syndrome occurs alone or is accompanied by additional neurologic or systemic abnormalities. Analysis of 11 recently discovered HSP genes provides insight into HSP pathogenesis. Hereditary spastic paraplegia is a clinical diagnosis for which laboratory confirmation is

sometimes possible, and careful exclusion of alternate and co-existing disorders is an important element in HSP diagnosis.

Treatment for HSP is presently limited to symptomatic reduction of muscle spasticity, reduction in urinary urgency, and strength and gait improvement through physical therapy. Prenatal genetic testing in HSP is possible for some individuals with the increasing availability of HSP gene analysis.

**Identification and management of intrathecal baclofen pump complications: a comparison of pediatric and adult patients.**

Vender JR, Hester S, Waller JL, Rekito A, Lee MR. Department of Neurosurgery, Medical College of Georgia, Augusta, Georgia 30912, USA. [jvender@mail.mcg.edu](mailto:jvender@mail.mcg.edu)

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refractory spasticity. The catheter, pump, and wound are subject to numerous complications both at the time of implantation and throughout the life of the implanted system. Careful technique, close observation, and aggressive evaluation and correction of problems can reduce the incidence and severity of the complications when they occur.

**The natural history of Primary Lateral Sclerosis.** Gordon PH, Cheng B, Katz IB, Pinto M, Hays AP,

SOURCE: J Neurosurg. 2006 Jan;104(1 Suppl):9- 15.

<http://tinyurl.com/ka28w> OBJECT: Intrathecal baclofen therapy is an effective means of treating intractable spasticity and dystonia in the pediatric and adult population. The authors present a review of complications encountered in a series of 314 pump and catheter-related procedures. Overall, there was a statistically significantly higher percentage of procedures for overall complication management and wound complication management in pediatric patients compared with adult patients. CONCLUSIONS: Intrathecal baclofen therapy is a highly effective treatment option for patients with medically

Mitsumoto H, Rowland LP. Department of Neurology, Columbia University, New York, NY, USA. [PHG8@columbia.edu](mailto:PHG8@columbia.edu) SOURCE: Neurology. 2006 Mar 14;66(5):647-53. <http://tinyurl.com/qIs49> OBJECTIVE: To define the syndrome of PLS and disorders that contain features of both ALS and PLS, to determine the time beyond which PLS is less likely to become ALS clinically, and to determine the outcome of people with PLS and those who develop lower motor neuron (LMN) signs. Dr. Gordon

(who, incidentally, is Mark Weber's neurologist) divides what we all call PLS into 4 diagnostic categories: 1. Autopsy proven PLS - clinically diagnosed PLS with no loss of lower motor neurons shown at autopsy 2. Clinically pure PLS - evident upper motor neuron signs, no focal muscle atrophy and no denervation in EMG 4 years from symptom onset. Age at onset after 40. 3. Upper motor neuron (UMN) dominant ALS - symptoms less than 4 years, or disability due predominantly to UMN signs but with minor EMG denervation or lower motor neuron signs on examination, not sufficient to meet diagnostic criteria for ALS. 4. PLS Plus - Predominantly UMN signs but also clinical, laboratory, or pathological evidence of dementia, parkinsonism, or sensory tract abnormalities.

(This article is one of several that resulted from the PLS Symposium that was held in June, 2004 in San Diego.)

### **Evidence for central abnormality in respiratory control in Primary Lateral Sclerosis**

Gouveia RG, Pinto A, Evangelista T, Atalaia A, Conceicao I, de Carvalho M. Department of Neurology, Neuromuscular Unit, Hospital de Santa Maria. Amyotroph Lateral Scler Other Motor Neuron Disord. 2006 Mar;7(1):57-60.

<http://tinyurl.com/q4gbb> Six patients

meeting previously proposed diagnostic criteria for PLS were submitted to a number of respiratory tests. Our results show that the diaphragm is not affected in this condition, but some respiratory function tests (RFT) and PNO had abnormal values. Voluntary muscular activation to perform RFT may be limited in these patients. PNO and polysomnography suggest that respiratory central drive dysfunction can occur when upper motor neurons are severely affected, in PLS. However, we did not verify progression on follow-up.

**Movement-related cortical potentials in PLS.** Bai O, Vorbach S, Hallett M, Floeter MK. Human Motor Control, National Institute of Neurological Disorders and Stroke, National Institutes of Health, Bethesda, MD. Ann Neurol. 2006 Mar 24;59(4):682-690 [Epub ahead of print] <http://tinyurl.com/hwlwe> Some patients with PLS have a clinical course suggestive of a length-dependent dying-back of corticospinal axons. We measured movement-related cortical potentials (MRCPs) in these patients to determine whether cortical functions that are generated through short, intracortical connections were preserved when functions conducted by longer corticospinal projections were impaired.

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MRCPs produced by finger taps were markedly reduced in PLS patients, including components generated by premotor areas of the cortex as well as the primary motor cortex. In contrast, the beta-band event-related desynchronization from the motor cortex was preserved. These findings suggest that impairment in PLS is not limited to the distal axons of corticospinal neurons, but also affects neurons within the primary motor cortex and premotor cortical areas. The loss of the MRCP may serve as a useful marker of upper motor neuron dysfunction. Preservation

of event-related

desynchronization suggests that the cells of origin differ from the large pyramidal cells that generate the MRCP.

### **Synapse**

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**EVENT PHOTOS Spring Fling, April 2-4, 2006 Berkeley Springs, WVA**

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Bettie Entire Group at Spring Fling Ronnie Grove, hostess of Spring

Jo and Don Wilson with Fling, being accosted by Pirates

Sue Me **PLSers on Vacation**

The Linda Neilsen on Great Wall of China Folks in standing photo (from the

Gentner's and Campbell's in left) Bride's father (Ronghai) , Derek Neilson (Groom), Zhao Wenxin (or  
Virgin Gorda, BVI

Vicky: Bride) Linda Neilson (me), Dennis Neilson, Bride's uncle, Bride's mother (Huawei) **SP**  
**Foundation Board Meeting**

Jean, Malin, Paul & Kris Frank, Mark & Linda Lenore & Jim  
Larry, Michelle & Karen Craig & Linda