

Synapse

Volume 14, Issue 1

Winter, 2011

Newsletter of the Spastic Paraplegia Foundation

Memorial Garden

a winter pick-me-up

see article p. 16



President's Letter:

Spring is coming...



Mike Podanoffsky
SPF President

I am writing this in late November but you'll read it in late January. The holidays and the New Year are a time for family and reflection. I hope yours brought you joy, comfort and rest. Spring is around the corner.

The past several months we have been bringing together a lot of medical researchers to discuss our conditions. They are all very eager

to renew their attention to motor neuron diseases. I want to share with you some of their efforts and thinking.

There is an ongoing project at Harvard (*and maybe other institutions*) to grow nerve cells. What they want to grow isn't a perfect nerve cell – that has been done. What they literally want to do is to take cells (*for instance, skin cells*) from someone with an upper motor neuron disorder like HSP (*or PLS*) and reprogram them to grow into nerve cells. This will reveal how the resulting neuron malfunctions. The goal is to then be able to test possible corrective measures.

There is research being conducted at The University of Massachusetts to block faulty nerve cells through a natural mechanism called RNAi. When cells divide, DNA produces RNA, which serves as a template needed to make proteins that cells need to differentiate, that is, to become a nerve cell instead of some other cell. When there is a mutation, the RNA produced is also faulty. One strategy using RNAi aims to block these faults.

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Synapse

Volume 14, Issue 1 - Winter 2011

The Spastic Paraplegia Foundation Inc. (SPF) is a national, not-for-profit, voluntary organization. It is the only organization in the Americas dedicated to Primary Lateral Sclerosis (PLS) and Hereditary Spastic Paraplegia (HSP).

Synapse Editors

Thurza Campbell Senior Editor
Sarah Roberts-Witt Events
Jim Campbell..... Caring

Published four times a year for the HSP/PLS community Available online at www.sp-foundation.org

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SPF Board of Directors:

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SPF Medical Advisor:

John K. Fink, M.D., University of Michigan

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SPF News from the Board

Medical Institutions that Welcome PLS and HSP Patients

We have expanded the list of clinics that have indicated they would welcome additional PLS and/or HSP patients. We now have a total of 51 NEALS institutions plus one other institution that welcomes new PLS patients. Thirty eight of these clinics are also welcoming HSP patients. The chart also includes the name of a physician who has agreed to be a point of contact and a phone number for making an appointment at the clinic. If you are not already being seen by a neurologist, we encourage you to contact the listed institution nearest you and arrange for an initial appointment. Please contact Jim Campbell by phone 508-653-5246 or email jimthurza@comcast.net if you have any difficulty arranging an appointment using the contact phone number listed. See complete listing pp 6-7.

NEALS/SPF Collaboration Working Group Meeting

Submitted by Jim Campbell

The first meeting of the group working was a full day conference held at Massachusetts General Hospital on November 11, 2010 to establish collaboration between SPF and NEALS. The conference was funded by a generous donation from a member of the SPF Board who wishes to remain anonymous. In attendance were the following neurologists from medical institutions across the US:

<u>Institution</u>	<u>Neurologists</u>
Massachusetts General Hospital	Steve Han Merit Cudkowicz, Nazem Atassi, James Berry,
University of California, SF	Jeffrey Rosenfeld Catherine Lomen-Hoerth
University of Michigan	John Fink
Northwestern University	Teepu Siddique
Hospital for Special Care	Jinsy Andrews
Baystate Medical Center	Carmel Amon
Drexel University	Terry Heiman-Patterson
Albany Medical Center	James Wymer
University of Kansas	Mazen Dimachkie

Barrow Neurological InstituteShafeeq Ladha
Also in attendance were SPF President Mike Podanoffsky and Board members Corey Braastad and Jim Campbell. A summary of the various elements of the meeting follows:

Current State of Research in PLS/HSP

Dr. Kevin Eggan, a PhD from Harvard University, summarized his work using skin cells of ALS patients to create induced pluripotent stem cells (iPS) and then lower motor neurons and most recently upper motor neurons. He noted the motor neurons have the potential for studying disorders and evaluating therapies much more efficiently than the mouse models of the past. His lab is already using iPS motor neurons to screen drugs for ALS.

Dr. John Fink spoke of the advanced state of gene identification for HSP and relatively little progress on the PLS front. He described a natural history (progress of symptoms in a patient over time) protocol he is establishing using standardized measures of outcome. He stated that PLS may not be a single disorder but a syndrome with clinical overlap with progressive spastic paraparesis in HSP, PLS and some ALS patients.

Dr. Nazem Atassi summarized the numbers of drugs in clinical trial for ALS patients. Citing www.clinicaltrials.gov he noted there were currently 150 ALS trials of which 94 were interventional and the remaining only observational. By comparison he was aware of only 8 clinical studies for PLS and HSP all of which were observational.

Identifying Ways To Increase PLS Research

Jim Campbell summarized the current SPF Research Grant evaluation process which includes the stated goal of annually allocating 50% of available research funds to each of the two disorders HSP and PLS. This year SPF received 18 HSP proposals, 3 joint proposals and only 1 PLS proposal. After receiving the assessment from its Scientific Advisory Board, the SPF Board voted to fund 2 HSP proposals, 1 joint proposal and 1 PLS proposal. *(The odds of successful PLS research proposals were not lost on researchers present.)*

To most effectively promote more PLS research the consensus of the group discussion was that a second PLS Research Conference was needed. The most recent prior conference was in 2004 organized by the Siddiques with seed money provided by four PLS families. Out of this conference hopefully would come consensus on where the research gaps are and where we need to go.

Clinical Care of PLS/HSP

Dr. Cathy Lomen-Hoerth described a PLS clinical trial she is conducting to measure the effect of the intrathecal baclofen pump with before and after comparison of patients at multiple centers. She has noted an improvement in the gait function and even oral function in some PLS patients.

Dr. Jinsy Andrews summarized some of the results from a patient needs study of the SPF community she and Dr. Nazem Atassi authored. (A preliminary report summarizing their findings can be found on pages 4 and 5 of this issue of *Synapse*.)

Current Registry Efforts

Dr. Teepu Siddique summarized his PLS registry effort at Northwestern University. He said he started his registry in the mid 90's with strict attention to diagnosis verification and follow up ascertainment to be certain the diagnosis was still valid. He said the registry contains blood samples of 190 PLS patients with 9 of them also providing corticospinal fluid (CSF) samples. He said a complete registry should ideally have the following: Patient blood samples as well as samples from parents and siblings, family history, patient clinical history, patient skin samples, environment history questionnaire and for deceased patients, brain and spinal cord autopsy.

Dr. James Berry reported on the current MGH-led NEALS study to collect biomarker samples from ALS patients as well as PLS and HSP patients and controls. He said the NEALS consortium currently has blood samples from 190 ALS patients, 16 PLS patients and 11 HSP patients. NEALS has used its consortium network to create a sample repository where the biomarker samples are currently collected at 30 institutions, but can be accessed by NEALS consortium members after review by a control committee.

Alex Sherman reported on the electronic data systems NEALS uses to capture data, manage the storage and dissemination of data and samples and manage the trial data collection for its ALS Registry. He said that NEALS currently has 36,000 samples from ALS patients and that the cost of sample storage needed to be considered in the establishment of any new registry. He emphasized the strong degree of protection NEALS offers in terms of separating identity of patients from their stored data and samples.

Registry Group Discussion

The consensus regarding registries was that more work is needed particularly in the area of collecting natural history data from patients. Natural histories with standardized measurement of symptoms would be particularly helpful when it comes to enrolling individual patients in clinical trials. With accompanying natural history data the efficacy of a therapy could be more precisely calibrated. In addition it was noted that while there are multiple PLS registries there is no central location at which information on the type and quantity of data and samples stored is available, so perhaps NEALS and SPF would have a role to play here.

Conclusion of Conference

A number of neurologists expressed the feeling that the day was worthwhile in terms of education on prior and current research initiatives and building a team spirit to ramp up the efforts to better serve the PLS and HSP community. Many expressed the willingness to participate in a PLS research conference. The NEALS co-leaders and SPF representatives agreed to define the next steps the collaboration should take in the near future.

Building the Path to Better Care and Treatment

*Submitted by Dr. Jinsy Andrews
(Hospital for Special Care)*

Earlier this year, SPF surveyed patients via their website to gather statistics on patient symptoms and the treatment they are receiving with the anticipation that this information will help create a foundation on improving clinical care and potential development of clinical trials.

A total of 221 people responded to the survey and majority of them were confirmed as having Hereditary Spastic Paraparesis (HSP) or Primary Lateral Sclerosis (PLS). Interestingly, more than 50% of people with PLS and HSP had to see more than 2 physicians to establish their diagnosis (approximately 20% had to see more than 5) suggesting a need for creating a network of specialized care. (*Fig 1*). Part of the diagnostic process for HSP is determining family history as it is a genetic disorder. Despite commercial availability of the test, more than half of patients with

HSP did not have genetic testing. The data also revealed that the most bothersome symptom for both PLS and HSP was walking difficulty and spasticity which may be a potential symptom to target therapy for clinical trials. The survey also showed that many patients do not go to a multidisciplinary clinic (*a clinic where there is physical therapy, occupational therapy, speech therapy, etc.*). This is a subject for concern considering that these diseases can affect multiple body systems (Fig. 2).

In a separate survey of NEALS institutions, the number of PLS and HSP patients seen at each institution

was recorded. Fig. 3 shows that a large number of NEALS institutions see significant numbers of both PLS and HSP patients, thus underscoring the logical collaboration between NEALS and SPF. Preliminary data from the survey and the NEALS/SPF collaboration has created a platform from which we can better address the needs of people living with these diseases and develop priorities for research. Additional data from the SPF community survey is being prepared for submission to a medical journal. Thanks to all who responded to the SPF community survey. Your participation gives us valuable insight into improving patient treatment.

Figure 1: The number of doctors visited to establish diagnosis

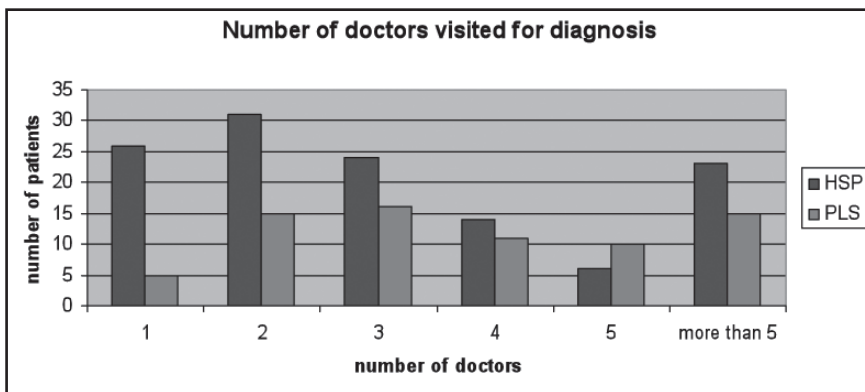


Figure 2: The number of patients that go to a multidisciplinary clinic

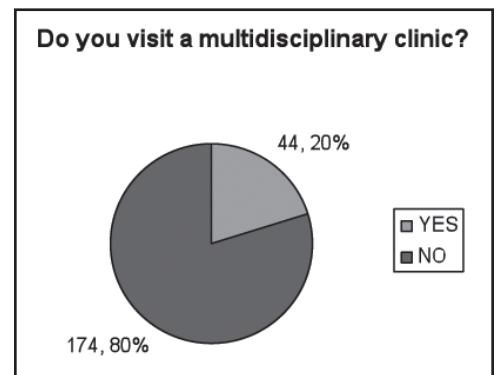
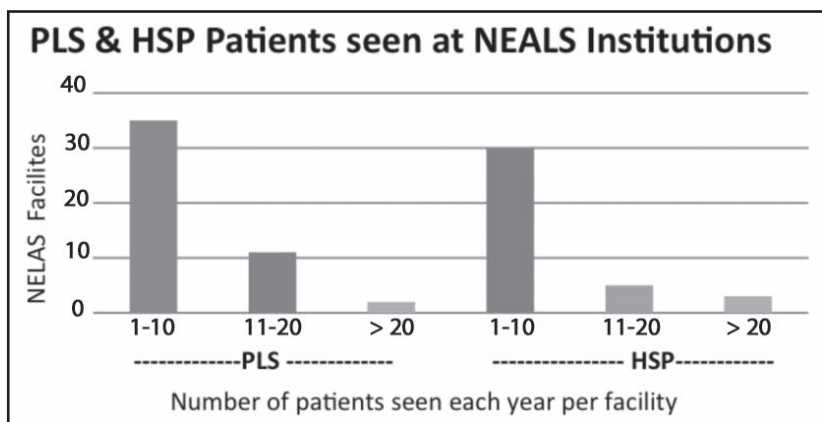


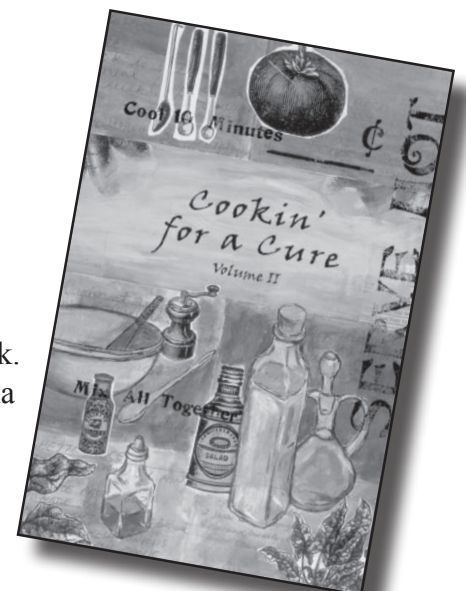
Figure 3: PLS/HSP patients seen at NEALS Institutions each year



Cookin' for a Cure II is Hot Off The Press

Thanks to Natasha Schaff for chairing our second *Cookin' for a Cure* cookbook. Natasha wanted to do a fund raiser in honor of her father who has HSP. Natasha started this project in May, 2010.

Order via the SPF website www.sp-foundation.org/store.htm or mail a \$13.00 US check made payable to SPF to: Linda Gentner 1605 Goularte Place, Fremont, CA 94539 (The price includes \$3.00 for shipping.)



NEALS Hospitals & Clinics Treating PLS and HSP

<p>University of Arkansas <i>Stacy Rudnicki</i> Little Rock, AR 72205 501-686-5838</p>	<p>PLS/HSP</p>	<p>Emory <i>Michael Benatar</i> Atlanta, GA 30322 404-778-3444</p>	<p>PLS/HSP</p>	<p>Baystate Medical Center <i>Carmel Armon</i> Springfield, MA 01199 413-794-1150</p>	<p>PLS</p>
<p>Barrow Neurological Institute <i>Shafeeq Ladha</i> 602-406-6262 Phoenix, AZ 85013 602-406-6262</p>	<p>PLS/HSP</p>	<p>University of Iowa Hospitals and Clinics <i>Andrea Swenson</i> Iowa City, IA 52242 319-356-4505</p>	<p>PLS</p>	<p>Johns Hopkins <i>Nicholas Maragakis</i> Baltimore, MD 21287 410-614-9874</p>	<p>PLS/HSP</p>
<p>Phoenix Neurological Associates <i>David Saperstein</i> Phoenix, AZ 85018 602-258-3354</p>	<p>PLS/HSP</p>	<p>Northwestern University* <i>Teepu Siddique</i> Chicago, IL 60611 312-695-0709</p>	<p>PLS/HSP</p>	<p>University of Michigan <i>Kirsten Gruis</i> Ann Arbor, MI 48109 734-936-9020</p>	<p>PLS/HSP</p>
<p>Phoenix Neurological Associates <i>Todd Levine</i> Phoenix, AZ 85018 602-258-3354</p>	<p>PLS/HSP</p>	<p>University of Chicago - <i>Kourosh Rezania</i> Chicago, IL 60637 773-702-9200</p>	<p>PLS/HSP</p>	<p>Saint Mary's Healthcare / Michigan State University <i>Deborah Gelinas</i> Grand Rapids, MI 49503 616-493-9727</p>	<p>PLS/HSP</p>
<p>University of British Columbia <i>Hannah Briemberg</i> Vancouver, BC, CAN, V5Z 1M9 604-822-7929</p>	<p>PLS</p>	<p>University of Chicago <i>Raymond P. Roos</i> Chicago, IL 60637 773-702-5659</p>	<p>PLS/HSP</p>	<p>St. Louis University <i>Ghazala Hayat</i> St. Louis, MO 63104 314-977-6086</p>	<p>PLS/HSP</p>
<p>University of California, Irvine <i>Tahseen Mozaffar</i> Orange, CA 92868 714-456-2332</p>	<p>PPLS/HSP</p>	<p>University of Kansas Medical Center - <i>Mazen Dimachkie</i> Kansas City, KS 66160 913-588-0039</p>	<p>PLS/HSP</p>	<p>Carolinas Medical Center - <i>Benjamin Rix Brooks</i> Charlotte, NC 28203 704-355-2000</p>	<p>PLS/HSP</p>
<p>University of California, Davis, Medical Center - <i>Bjorn Oskarsson</i> Sacramento, CA 95817 916-734-3588</p>	<p>PLS/HSP</p>	<p>Beth Israel Deaconess Medical Center - <i>Elizabeth Raynor</i> Boston, MA 02215 617-667-8130</p>	<p>PLS</p>	<p>Wake Forest University School of Medicine <i>Michael Cartwright</i> Winston-Salem, NC 27157 336-716-4344</p>	<p>PLS/HSP</p>
<p>University of California, San Francisco <i>Catherine Lomen-Hoerth</i> San Francisco, CA 94143 415-353-2122</p>	<p>PLS</p>	<p>Brigham And Women's Hospital <i>Anthony Amato</i> Boston, MA 02215 617-732-8046</p>	<p>PLS/HSP</p>	<p>Duke University <i>Richard Bedlack</i> Durham, NC 27707 919-668-7600</p>	<p>PLS/HSP</p>
<p>University of California, Fresno <i>Jeffrey Rosenfeld</i> Fresno, CA 93701 559-264-9100</p>	<p>PLS/HSP</p>	<p>University of Massachusetts Medical Center - <i>Johnny Salameh</i> Worcester, MA 01655 508-334-9040</p>	<p>PLS/HSP</p>	<p>Dartmouth-Hitchcock Medical Center <i>Elijah Stommel</i> Lebanon, NH 03756 603-650-5104</p>	<p>PLS/HSP</p>
<p>Hospital for Special Care <i>Jinsy Andrews</i> New Britain, CT, 06053 860-827-4924</p>	<p>PLS/HSP</p>	<p>Massachusetts General Hospital <i>Nazem Atassi</i> Boston, MA 02129 617-726-1728</p>	<p>PLS/HSP</p>	<p>Robert Wood Johnson Medical School <i>Jerry Belsh</i> New Brunswick, NJ 08901 732-235-7733</p>	<p>PLS</p>
<p>Mayo Clinic, Florida <i>Kevin Boylan</i> Jacksonville, FL 32224 904-953-6915</p>	<p>PLS/HSP</p>	<p>Lahey Clinic <i>Jim Russell</i> Burlington, MA 01805 781-744-7273</p>	<p>PLS/HSP</p>		

University of Nevada School of Medicine <i>David L. Ginsburg</i> Las Vegas, NV 89102 702-671-5025	PLS/HSP	Oregon Health & Science University <i>Jau-Shin Lou</i> Portland, OR 97239 503-494-7772	PLS	University of Washington <i>Michael Weiss</i> Seattle, WA 98195 206-598-7688	PLS
Upstate Neurology <i>James Wymer</i> Albany, NY 12205 518-438-0922	PLS	Penn State Hershey <i>Zachary Simmons</i> Hershey, PA 17033 800-292-332	PLS/HSP	Dean Health System <i>Oliver Ni</i> Madison, WI 53719 608-252-8000	PLS/HSP
Beth Israel Medical Center Albert Einstein College - New York - <i>Stephen Scelsa</i> New York, NY 10003 212-844-8490	PLS	University of Pittsburgh <i>Sasa Zivkovic</i> Pittsburgh, PA 15213 412-692-4917	PLS/HSP	Medical College of Wisconsin <i>PE Barkhaus</i> Milwaukee, WI 53226 414-805-5224	PLS/HSP
Columbia University - MDA/ALS Center <i>Hiroshi Mitsumoto</i> New York, NY 10032 212-305-1319	PLS	University of Pittsburgh <i>David Lacomis</i> Pittsburgh, PA 15213 412-692-4917	PLS	WVU School of Medicine <i>Laurie Gutmann</i> Morgantown, WV 26508 304-598-4800	PLS/HSP
SUNY Upstate Medical University <i>Jeremy Shefner</i> Syracuse, NY 13210 315-464-4243	PLS/HSP	Drexel University College of Medicine <i>Terry Heiman-Patterson</i> Philadelphia, PA 19103 215-762-6915	PLS/HSP	<i>* Not within NEALS consortium</i>	
Cleveland Clinic <i>Rebecca Kuenzler</i> Cleveland, OH 44195 800-223-2273	PLS/HSP	Rhode Island Hospital, Brown University <i>George Sachs</i> Providence, RI 02903 401-444-8795	PLS/HSP		
Ohio State University <i>John T. Kissel</i> Columbus, OH 43210 614-293-4969	PLS	Texas Neurology <i>Daragh Heitzman</i> Dallas, TX 75214 212-827-3610	PLS/HSP		
Ohio State University <i>Yasushi Kisanuki</i> Columbus, OH 43210 614-293-4969	PLS/HSP	University of Texas Health Science Center, San Antonio <i>Carlayne Jackson</i> San Antonio, TX 78229 210-450-9700	PLS		
London Health Sciences Centre <i>Christen Shoesmith</i> London, Ont., CAN N6A 5A5 519-663-3597	PLS/HSP	University of Virginia <i>Lawrence H. Phillips, II</i> Charlottesville, VA 22908 434-924-0000	PLS		
University of Toronto <i>Lorne Zinman</i> Toronto, Ont., CAN M5S 1A1 416-480-4213	PLS	University of Vermont <i>Rup Tandan</i> Burlington, VT 05405 802-847-4589	PLS/HSP		

National Conference

FRIDAY, June 10, 2011

- 9:00 AM-2:00 PM Board Meeting
3:00-5:00 PM Registration
5:00-6:00 PM Hospitality Time/Greet and Meet
6:00-9:00 PM Dinner
Welcome – Mike Podanoffsky
Speaker from Binder and Binder-
“Disability and the Law”
NEALS Report

SATURDAY, June 11, 2011

- 8-9 AM Registration
12 Noon - 6 PM Vendors/Exhibitors Viewing
9 AM Start Time: Conference Room/Registration/
Vendors/Exhibitors 3rd floor
9 :00-9:15 Craig Gentner – Welcome and
Instructions for Conference
9:15 -9:45 Mark Weber – Grants
9:45-10:45 Cory Braastad, PhD- I hour –Basic
Genetics & Genetic Testing for HSP/PLS
10:45-11:00 Break
11:00-12:00 Dr. Fink- Research Update
12:00-12:15 Craig Gentner- Announce lunch, etc.
12:15- 1:30 Lunch- Dr. Jinsy Andrews – Treatments
for PLS and HSP
1:30-1:45 Break
1:45-2:45 Dr. Lyn Litchke – Chair Yoga
2:45-3:45 John Paul Liang – Acupressure and
Acupuncture Treatments
3:45 -4:05 Kathi Geisler – History of Spastic
Paraplegia Foundation
4:05 -4:30 Break
4:30 – 5:30 Breakout Sessions (Located on 2nd floor)
- Katie St. Mars - *The Benefits of Water:
Seen & Unseen Benefits of Aquatic
Therapy*
- Tom DiBello – *21st Century Orthotics
and WalkAide*
- Allie Keaton – *Service Dogs*
5:30 PM Saturday Session Ends – Dinner on Your
Own

SUNDAY, June 12, 2011

- 12 Noon Hotel Checkout, if leaving on Sunday

**2011 SPF Annual Conference:
“Rocketing Toward The Cure
Through Research”
Houston, TX June 10-12, 2011**
Hilton Americas- Houston
1600 Lamar, Houston, Texas, 77010
www.americashouston.hilton.com
Ashton Hecker, Chair. 832-453-0394

HILTON AMERICAS ONLINE ROOM

RESERVATIONS:POG's (*Personalized Online Group Page*) has been established for those that choose to register for hotel accommodations online.

The web page address for registering online for a 1SP Room (Non ADA room; kings and double bed rooms are available) is: http://www.hilton.com/en/hi/groups/personalized/H/HOUCVHH-1SP-20110610/index.jhtml?WT.mc_id=POG

The web page address for registering online for a 1SR Room (ADA room with roll in showers, no tub; all double bed rooms) is: http://www.hilton.com/en/hi/groups/personalized/H/HOUCVHH-1SR-20110610/index.jhtml?WT.mc_id=POG

The web page address for registering online for a 1SF Room (ADA room with tub only; kings and double beds available) is:
http://www.hilton.com/en/hi/groups/personalized/H/HOUCVHH-1SF-20110610/index.jhtml?WT.mc_id=POG

CALL IN RESERVATIONS

To call in directly to the hotel: 713-577-6667
Dialing this number, the guest will speak to one of 3 reservationists. It will not rollover to a call center. If someone does not answer the phone, they will go to voicemail to leave a call back number. One of 3 ladies will call back as soon as possible; Kamaria Price, Savannah Gomez, or Damita Wallace.

TRANSPORTATION: All Wheelchair Accessible

Super Shuttle 281-230-7275 www.supershuttle.com
They are located at baggage claim at the airports and attendees can go on-line to purchase tickets in advance.
Yellow Cab 713-236-1111

This is the only cab company with ramps for wheelchairs.

PARKING

Hilton Garage: Hotel garage has 4 handicap spots on each floor (*on the up side (entrance) and on the down (exit) side*) that are located next to the elevators.

So as you go up in the garage to the 7th level, you will see them next to the elevators and when you go down to the ground floor you will see them next to the elevators. There is a bar that has a height of 7ft as you enter the garage. Any van or truck that has a height higher than 7 feet will not fit into our garage.

AceParking: (1702½ Rusk Street) Has parking available for RVs, but does not have electricity for hookup.

Discovery Green: (*Across street from hotel*) 713-400-7336

Toyota Center: (1510 Polk Street) 713-758-7200

Street Parking: There is a charge at the meter until 6pm Monday – Saturday. After 6pm it is free to park on the street. Charge meters **are not** located on each parking space, but are spaced along the street.

HILTON AMERICAS ROOM REFRIGERATOR

Mini bar in each hotel room is available for refrigeration for medicines. Remember to ask for mini bar key upon check in. If the guest decides they need a refrigerator in addition to the mini bar, the charge is \$25.

CONFERENCE SPEAKERS

Dr. Jinsy Andrews

Dr. Jinsy Andrews is board certified in neurology, electrodiagnostic medicine and neuromuscular diseases. She completed a two year fellowship in neuromuscular disease at the Neurological Institute of Columbia University and now serves as the Director of Research and Clinical Trials at Hospital for Special Care in Connecticut. Dr. Andrews's interests include clinical research studies and clinical trials in neuromuscular diseases. She is co-chairing HSP/PLS Taskforce created through the SPF and NEALS collaboration to improve care and advance research in HSP and PLS.

Binder and Binder (*Individual speaker TBA*)

Binder & Binder® is America's Most Successful Social Security Disability Advocates®. With locations nationwide, we serve clients' Social Security Disability and Supplemental Security Income needs in all 50 states, Puerto Rico, the Virgin Islands and all U.S. territories. Binder & Binder® has offices in strategic locations all across the country. Our nationwide presence allows us to bring in an army of highly qualified advocates and top-level executives to handle SSD and SSI cases across the country.

Website: www.binderandbinder.com

Corey Braastad

Current

Manager, Laboratory Operations and Practical Process Improvement at Athena Diagnostics (ThermoFisher): <http://linkedin.com/pub/coreybraastad>

Board of Directors Member at Spastic Paraplegia Foundation

Education

University of Massachusetts Medical School Postdoctoral Fellowship, Cell Biology, 2002-2004

Brown Medical School Postdoctoral Fellowship, Cancer Biology, 2001-2002

Brown University Ph.D., Molecular and Cellular Biology and Biochemistry, 1996-2001

University of Massachusetts Dartmouth B.S., Biology, 1992-1996

Tom DiBello, CO, FAAOP

Bachelor's Degree in Pharmacy from Duquesne University, Pittsburgh, Pennsylvania

O&P certificate program at Northwestern University, Chicago, Illinois

Residency at the University of Oklahoma (UO), Oklahoma City

Tom DiBello has become one of O&P's most distinguished leaders. At the American Academy of Orthotists and Prosthetists (the Academy), he has served on the board of directors, as a member of its executive committee, and in 2001, as its president. Dynamic Orthotics and Prosthetics has three offices that employ ten full-time practitioners and about 15 technicians. <http://www.dynamicoandp.com>

John K. Fink, M.D.

Professor, Department of Neurology Director, Neurogenetic Disorders Program

University of Michigan, Ann Arbor

<http://www.med.umich.edu/geriatrics/research/directory/fink.htm>

Dr. Fink is the SPF Medical Advisor. In addition to being a one of the world's foremost investigators of upper motor neuron disorders, he also maintains the largest clinic in the U.S. for persons with HSP or PLS. After graduating in biology from University of Cincinnati, and Medical School at the Medical College of Ohio, he trained as a neurologist at the University of Virginia and in specialized aspects of neurology and medical genetics at the National Institute of Health. As a Professor of Neurology at the University of Michigan he directs the neurogenetic disorders program. He also studies genes that cause these disorders. He is a recipient of a Spastic Paraplegia Foundation research award in 2003 and again in 2006.

Kathi Geisler, a marketing-communications professional who helped develop the online HSP patient community in 1996 and organized the first HSP regional support group in 2000, is the co-founder of the Spastic Paraplegia Foundation, Inc. In 2001, she and SPF Board Member Mark Weber, with the guidance of SPF Medical Advisor John K. Fink, M.D., organized a Steering Committee comprised of 20 individuals from the HSP and PLS patient communities. The goal was to develop one foundation dedicated to both conditions and in February, 2002, the Spastic Paraplegia Foundation was incorporated. Kathi served as the SPF Vice President for its first three years, developing educational, fundraising and support programs as well as the SPF website.

Allie Keaton is the President and trainer for My Service Dog, Inc. which is a 501(c)(3) nonprofit organization established in 2004 to train Service Dogs, Assistance Dogs, and Hearing Dogs to promote independence and socialization for the disabled in and around, but not limited to, Houston, Conroe, and Woodlands areas. Allie commented to me: "My Service Dog, Inc. does not rescue dogs, but trains dogs to rescue people." www.myservicedog.com

John Paul Liang, M.S.O.M., L.Ac. received his B. S. from Cornell University and his M.S. in Oriental Medicine Degree from the American College of Acupuncture & Oriental Medicine. As the director of Planning and Development, he is in charge of growth and development at the American college of Acupuncture. Currently, he is a practitioner at the ACAOM outreach facility of Methodist Hospital. He is a Diplomat of the NCCAOM in acupuncture, herbology, and Oriental medicine, and a licensed acupuncturist in Texas. He is also a lecturer at Rice University. <http://www.acupuncturecarehouston.com>

Dr. Lyn Litchke, Assistant Professor of Therapeutic Recreation Texas State University, San Marcos, Texas

Education:

B.S. The Ohio State University; M.S. Texas State University; Ph.D. Texas State University

My overall research area of interest is in improving athletic performance for persons with disabilities. Recently my research efforts have focused on utilizing respiratory resistance training to enhance athletic performance, quality of life, and self-efficacy for wheelchair athletes. I am currently seeking an adaptive yoga certification to incorporate into my research and teaching.

Katherine O. St. Mars, PT, DPT graduated from Virginia Commonwealth University's Medical College of Virginia with a Doctor of Physical Therapy degree. Upon graduating, she began her career with sports medicine, post-operative, general orthopedic & neurologic patients in the outpatient setting where she enabled the innovative use of video-game systems to improve many injuries & dysfunctions in a plethora of patient populations. After moving to San Antonio, TX, Katie applied her experience to children with neurodevelopment & orthopedic diagnoses, using aquatic therapy to achieve amazing results in improved function.

Mark Weber is a founding board member, co-chaired the Foundation Steering Committee and completed his first term as SPF President. He is an attorney with eleven years experience as an Assistant Attorney General and Assistant District Attorney. Mark has been an active community leader since 1999, launching and managing a PLS email support group, serving as editor of a PLS newsletter and organizing fundraising initiatives. The following year, he became active in the HSP online community. Mark holds a BA in economics and psychology and a JD. He was admitted to the Massachusetts Bar in December 1986. He was diagnosed with PLS in 1997. Mark and his wife and two young boys live in Sherman, CT. Mark serves the SPF as Legal Counsel and also chairs the Research Committee.

SYNAPSE APPEAL

Synapse costs lots of money to print and mail, and we need your help to keep it going for another year. Please use the enclosed response envelope to make a donation. Every little bit helps.

Event Reports

New England Team Challenge Merrimac, NH September 11, 2010

Submitted by John Swain, Chair

On a beautiful New England day, approximately 45 New England Team Challenge attendees gathered in Merrimac, NH, with smiles on their faces. Our Team Challenge started with



NE Team Challenge Begins @ Budweiser



NE Team Challenge Group

socializing and a cook-out complete with hamburgers, hot dogs, and all the fixings. After lunch, we heard from SPF President Mike Podanoffsky regarding research and SPF community

updates. In addition, Kathi Geisler demonstrated her new Dashaway stand-up walker, which some of us tried out. The stables of the famous Anheuser Bush Clydesdales were our next stop; that was followed by a tour of the brewery itself.

This event raised more than \$36,000 dollars. It was put together with the help of committee members Jim and Thurza Campbell, Joel and Bobbie Seidman, Bob Swain, Judy Johnson, chairperson John Swain, and Laurie LeBlanc and family.



NE Team Challenge Chair, John Swain

The Magnificent Mile Race Raleigh, NC September 19, 2010

Submitted by Sarah Witt, Chair

A record 1,203 participants turned out for The Magnificent Mile Race (www.magmilerace.com). This year's attendance marked a 50% increase from 2009. In addition, more than \$50,000 was raised for the SPF. The fifth annual event included competitive, recreational, and kids' races, as well as family friendly activities. It

also served as the North Carolina-USATF one-mile championship race. Bobby Mack, a former cross country All-American and all-conference runner at North Carolina State University, won the men's



Cece Russell once again turns in a winning performance



Sarah Witt and family joined by Carolina RailHawks mascot, Swoops

overall with a time of 4:10. This tied the state record he set at last year's event. The women's overall winner was Angelina Blackmon, a 23-year-old Cary resident who finished in a time of 5:02.

The Magnificent Mile Race is the only NC Triangle-area program specifically dedicated to raising awareness of and research funds for Primary



This year's event had more than 1,200 participants

Lateral Sclerosis and Hereditary Spastic Paraplegia. To date, nearly 3,500 participants have helped raise more than \$200,000 to fund research.

The Drive "Fore" SPF Golf Outing Monroe Township, NJ Sept. 20, 2010

Submitted by James F. Brewi, Chair

A beautiful early fall day in Jamesburg, NJ, greeted the golfers who participated in the second annual Drive "Fore" SPF Outing honoring James F. Brewi. Travelers Insurance once again



Off and Running

hosted the event and opened it up to other interested parties. As a result, the turnout was larger, with 120 golfers and an additional 60 people joining the group for

the evening reception. Travelers' Chairman and CEO, Jay S. Fishman, numbered among them.



Jim Brewi

At the reception, Maureen Brewi presented an update for the family and gratefully acknowledged all the efforts and generous support. She gave a special thanks to Lisa Tepper, Travelers' Regional President, for her tremendous leadership and support of Jim. Jim then introduced Annette Lockwood, who gave a heartfelt overview of the SPF mission as well as our search for a cure.

Annette presented Mr. Fishman with a digital photo album of the day's events that had an engraved note of thanks from SPF. Mr. Fishman then announced that the Travelers Foundation would once again donate \$100,000 to SPF. The amount raised at this year's event far exceeded last year's total. We are extremely grateful to the friends and family who donated, as well as to Travelers and Mr. Fishman.



SPF Presentation to Jay Fishman, CEO and Chairman of Travelers Insurance

The Clifton Park Rotary Event Clifton Park, NY September 24-26, 2010

Submitted by Isabel Prescott

At Riverview Orchards, our small farm in Rexford, NY, harvest time is extremely busy. Crowds of people come to pick apples, take hayrides, enjoy apple cider and donuts, and indulge in lots of fun on the farm. The harvest also provides an opportunity to hold events and fundraisers, which is what occurred on the weekend of September 24-26. The draw of autumn events on the farm, as well as functions sponsored by the local Rotary Club, brought out thousands of people.



Among the activities for visitors were four for which the money raised went entirely to the SPF. These included two pie-eating contests, one for kids and one for adults; a Corn Hole Tournament; a quilt raffle organized by a local women's group called Q.U.I.L.T.; as well as a Three-Dimensional Cookie Demonstration and sale orchestrated by my daughter-in-law, Jen. These events raised a total of \$500 for SPF, with the knowledge that each and every dollar counts for research toward a cure.

NY TeamWalk Mt Kisco, NY

September 25, 2010

Submitted by Ann Lakin, Chair

Our NY TeamWalk was held in Mt. Kisco on a beautiful fall day. About 75 people participated. We gathered mid-morning, then ate and socialized for an hour or so. Those who registered received



an SPF t-shirt as well as a goodie bag donated by local banks. Around noon, our group walked to the end of the street before circling back to our original destination. Afterwards, we enjoyed raffles from



local businesses. We had a very pleasant day marked by great weather, great fellowship, and great fund-raising. A special thanks goes to Meredith Gattuso, who brought 35 family members with her.



Meredith Gattuso and Ann Lakin

California TeamWalk & Connection Pleasanton, CA

October 1-2, 2010

Submitted by Linda Gentner



At our seventh annual California TeamWalk & Connection, approximately 80 people congregated on a picture-perfect weekend to raise awareness

and funds. Friday evening began with dinner at the Hilton, which was followed by dessert and a "share and compare" session. We had a "new" lady this year named Connie Crawford (HSP). She and her family drove from the Palm Springs area, which is more than seven hours away, to join us. Her daughter had recently found us on the Internet. It was an emotional experience for Connie to discover that she is not alone in her struggles.



Russ Ritchell of Independence Options makes his Be Safe at Home presentation

and my special Orange Glazed Breakfast Buns. Russ Ritchell of Independence Option gave the first presentation of the day entitled "Be Safe at Home". In addition to demonstrating the emergency medical alert system, Russ explained that fall detectors initiate an emergency call if a special "no movement" detector doesn't show movement that would be expected of the user. Then Ben Lambeth of Carolina Mobility and Seating demonstrated a stander, along with the latest power chairs from Pride and Sunrise Medical. I demonstrated the EasyPivot lift and the Alice lift from Apex.



Tony Beauchamp and Annette Lockwood try out Dashaways

After lunch, we were scheduled to have a "Seated Yoga" presentation courtesy of Alexis Gabard. Ms. Gabard was unable to attend, so she sent a CD for yoga style breathing and meditation. The last session of the day was a "Laugh Workshop" by Bob Clemmons. Everyone woke up to meet Tickle Me Elmo and generally laugh themselves silly.

Then came the grand finale -- the "SAWCAR 280" races for scooters and power chairs. Ronnie brought her hot four-wheeled scooter to defend her 2009 wins. No one else was driving a scooter, so Gary Fisher agreed to race her with his loaner scooter. However, the loaner didn't provide much competition and Ronnie easily took the two-lap race. Newcomer Kim Medlin drove her heart out to win the power chair race over five other drivers. Ronnie and Kim then raced for the "SAWCAR 280" Grand Champion title. Ronnie finished a chair length ahead of Kim and defended her title for the second year.

Those who didn't return home on Saturday gathered to share an evening meal, followed by chatting in the hospitality suite. All in all, the weekend was outstanding. Bettie Jo and I hope that everyone enjoyed the gathering as much as we did, and thanks to everyone who made the effort to attend.

Chasing a Cure Connection

Fairfax, VA

October 30, 2010

Submitted by Beth Anne Shultz, co-chair

The 'Chasing A Cure' connection event held in Fairfax, Virginia in late October of this year brought together five talented and informative professionals and an audience of sixty-one. This SPF gathering included a diverse group of people of all ages, some of whom traveled quite a distance in order to attend. In fact, eight states and two countries were represented.



Dr. Mary Kay Floeter chats with Maureen Brewi



Dr. John Fink and Beth Anne Shultz

Dr. Mary Kay Floeter and Dr. John Fink provided the latest facts and figures concerning PLS and HSP. Dr. Simon Fishman gave a lively and detailed presentation with regards to managing spasticity.

Jenna Justin, a physical therapist who has an interest in working specifically with clients suffering from neurological conditions, provided a particularly well-received tutorial about the importance of maintaining and improving motion and mobility. Leslie Horton, RN, concluded the day with a thought-provoking presentation entitled, "Is a Service Animal for You?". Regardless of the complexity of the issue, several of us thoroughly enjoyed the opportunity to see one of her four-legged companions in action. Numerous breaks and a long, relaxing lunch provided ample opportunity to meet and mingle with other guests.



Leslie Horton, RN, and her service dog

Tennessee TeamWalk & Connection

Nashville, TN

November 6, 2010

Submitted by Jim Sheorn

This year's event was held at Beyond Therapy, which specializes in physical therapy for those who have neurological disorders. We had a small group of regular attendees as well as a new couple. Jenna Briggs, DPT, was our guest speaker. She



Living with HSP/PLS

2 Those Who Care: Don Wilson

By Sarah Hughes



Don Wilson

Caring for a loved one changed the way one Triad man felt about helping others. Our “2 Those Who Care” recipient for this month is Don Wilson.

Wilson’s list of volunteer efforts is at least a mile long because he spreads himself over a large number of causes. The one that gets a significant amount of his attention is helping people with neurological diseases.

Don’s wife was diagnosed with Primary Lateral Sclerosis and he is her caregiver. Each year, Don plans a gathering of people with these diseases as a way to give them a chance to learn from each other. It is a way to share their successes and failures.

Don calls the day-long meeting a fun way to give out information that makes a difference.

(Ed. Note: The event mentioned is Autumn in Carolina)

The Memorial Garden Website

By Di Montague Jackson, Alberta Canada
www.mndmemorialgarden.org

Forever Friends Memorial Garden came into being in July of 2008 in RED DEER, ALBERTA, Canada. Supported and encouraged by many members of ‘PLS-FRIENDS’ and HSP support websites, as well as the local ALS Society, both with ideas and donations. The ‘Garden’ began with a few small bushes and a rockery in our front garden to remember some friends from the PLS Support site who had passed on. Since then many people across the world have sent photos of loved ones who have passed on with a motor neuron disease (PLS, HSP, ALS, etc.). Many also sent small amounts of soil from their loved one’s garden, seashells, plaques, butterfly ornaments along with donations of various sizes.

As I look out over ‘The Garden’ today, it has grown 20 fold since those early days. The various flower beds, bushes and shrubs are blooming! We have made special ‘photo-plaques’ of those being memorialized and placed them among the blooms. As you stroll

through the garden you will come upon many statues, both donated and purchased by us, that add a peaceful, spiritual feel. Truly, our loved ones and friends are remembered in a most beautiful way!

The picture on the cover is of the garden. Please go to the website to see more pictures, and to see how you can become a part of the memorial garden project.

Exercise And You: Why Stretch?

Contributed by Liz Wroblewski, MPT

We are often advised to do “stretching exercises” for spasticity management in PLS and HSP. Keep in mind that there is no definitive clinical research that correlates consistent stretching with an overall long term decrease in spasticity. Just what are we doing when we incorporate stretching into our daily routine?

Stretching is essential to maintain the pliability and length of the soft tissue surrounding a joint. This soft tissue includes the joint capsule, muscles, tendons, ligaments, fascia, nerves and blood vessels. If left in a shortened position, they will adapt to this shortened position and then limit the range of motion in the joint. Limited range can result in functional difficulties. For example, in prolonged sitting the hip flexors as well as the surrounding structures shorten restricting the hip extension we need to stand erect. If this range is not available in standing we then lose the optimal power of the gluteal muscles to support us and propel us forward in walking. In order to stand upright with tight hip flexors we may then over-arch the low back to maintain the trunk erect. This can be a cause of back pain. Another example would be shortening of the calf muscles with restriction of the ankle in dorsiflexion range. We need 10 degrees of dorsiflexion for level walking. If that amount is not available, standing balance is affected. Lack of range of motion of the ankle may be a determining factor in achieving less than optimal outcomes with the WalkAide or AFO.

Muscles have specialized bundles of tissue called the muscle spindle. This muscle spindle responds to stretch, i.e., when the spindle is stretched there is a reflex arc through the spinal cord to contract the muscle containing that spindle. The spindle responds to the velocity of the stretch as well as the change in

length of the muscle. Walking is a perfect example of how we “stretch” our muscle spindles. When we step forward with a normal stride length and heel strike, the hamstring and calf muscles are stretched. **If the muscles and soft tissue have shortened through lack of stretching and disuse, then the stretch stimulus to the shortened spindle engages that much sooner creating more tone in the muscle.** Also, the faster we attempt to walk, the more difficult it can become because of spindle sensitivity to increased velocity. Normally the increased tone would be modulated by inhibitory impulses at the spinal cord. We lack this inhibition due to corticospinal impairment. Our excitatory impulses predominate. This is the source of our spasticity or hypertonia. We lack reciprocal inhibition which would otherwise allow us to move in a smooth coordinated fashion, i.e. as one muscle group contracts (excitatory) the opposing muscle group would normally relax (inhibitory). Over time the unstretched spastic muscle loses length through loss of its contractile units called sarcomeres. This weakens the muscle and allow it to become overly sensitive to stretch. This sets up further shortening due to muscle spindle excitement resulting in the muscle contraction itself. It is potentially an endless loop of contract-tighter-contract to eventual contracture.

I hope I have conveyed well enough two reasons to systematically stretch in upper motor neuron syndromes that present with spasticity.

Fifth-grader with HSP Rode in the Hudson Kids Ride Pan-Mass Challenge

*Excerpted from an article by Jeff Malachowski/
Daily News staff The
MetroWest Daily News*

Owen Anketell, a fifth-grader with HSP rode his specially made three-wheel bicycle, which he pedals with his hands, along with several other kids during the 2nd annual Pan-Massachusetts Challenge Hudson Kids Ride. The event aims to help raise money for cancer research and to give back to the community. “I just thought it was cool and would be cool to help other people,” he said. When Anketell isn’t riding his bike around Hudson, he said he enjoys playing basketball, going horseback



riding, kayaking, skiing and waterskiing. Anketell said his favorite activities are skiing and waterskiing, despite making his mother nervous the first time he tried. “It feels good that I can give back to people,” he said. “People help me a lot, so I decided I should do something to help people.”

European Federation of HSP

Contributed by Fernando Gonzalez, President
<https://sites.google.com/site/eurohsp/home>

The European Federation of HSP has begun! We have been working since July of 2009 and our short name is EURO-HSP. The HSP groups from Denmark, France, Italy, Norway, Spain and the United Kingdom decided to create this Federation with the help of EURORDIS, the European coordinator of Patients with Rare Diseases.

EURO-HSP is an international apolitical, non confessional and non-profit association registered in Paris on July 2010 with the following objectives:

1. Keeping up with and supporting medical research on HSP and related conditions;
2. Central processing of information on such research and passing it on to the members of EURO-HSP
3. Promoting and improving contacts between practitioners concerned and persons affected by HSP and other related conditions;
4. Investigating social, political and cultural matters connected with the welfare of people affected by HSP and related conditions, as well as promoting and improving the exchange of information on such matters;
5. Promoting co-operation on an international level between national associations for people affected by HSP and related conditions;
6. Co-operating with or being members of other national and international organizations and institutions that have the objective of furthering the welfare of individuals affected by a physical disablement and, inter alia, those with a neuromuscular condition;
7. Gathering funds and collecting, managing, using and distributing contributions, legacies and donations or their benefits, for the purpose of stimulating and promoting both medical research into HSP and related conditions and the welfare of people affected by them.

8. Serving as a meeting point for any and all implicated groups (people with HSP, their families, investigators and scientists, doctors, nurses, health care workers, etc.) where all aspects of dealing with optimal maintenance of health can be sought, including alternative methods with rational basis.
9. Participating in media and media events in order to keep the general public as well as public and private institutions, European or International, aware of HSP issues and concerns.
10. Other activities not specifically contemplated in these statutes that may favor the promotion, support, and social and labor inclusion of people affected by HSP and / or related conditions.

If you would like to join us you will be more than welcome. If you are not European, if you are interested, you can join us as an Associate Member. You can see in our bylaws proposal (<https://sites.google.com/site/eurohsp/bylaws-proposal>), under Article II, section 1.2, that in spite of our name, you will have the same rights as Full Members except on European-specific matters.

Laughing Sickness: A Medical Mystery

By Anne Black Gray

Twenty-five-year-old Jessica Shephard always knows how to get a laugh. She's enjoying good health, friends and family when, without warning, she's struck by paralytic episodes. Doctors can't discover what's wrong, but laughter is mysteriously entangled in her illness. As relationships with friends, family and coworkers deteriorate and her life becomes threatened, she struggles to keep her job and independence.



...This work, admirably, shows the authorial intent to foster awareness of "orphan" diseases, the need for improved medical research and development of therapeutic approaches and a greater respect and understanding of those with disabilities...September 29, 2010

President Barack Obama
The White House
1600 Pennsylvania Ave N.W.
Washington, DC 20500
Re: Your humble act of kindness



Dear Mr. President,

A few days ago, you greeted a young lady using a walker to stand, Jackie Wellman. You may not be aware of how meaningful your action was, in helping her to stand, once her walker had been safeguarded by the Secret Service. As she told me in her e-mail,

"When I approached him the Secret Service took my walker. He reached out both his hands to grab mine and said, "Don't worry, I've got you."

She and I, and at least 30,000 others in the United States, suffer from a rare neurologic disorder, Hereditary Spastic Paraplegia, or a related disease, Primary Lateral Sclerosis, which eventually impair our ability to walk, and in some cases to speak and swallow normally. You are now also famous in our community as someone who respects and cares about "our people." I am on the national board of directors of the Spastic Paraplegia Foundation, and all of our members now know of your kindness, good will, and caring.

Thank you for caring for her and for everyone who needs help.

Sincerely,
Malin Dollinger, M.D.

Wheelchair Hoopster Pushes Herself To Succeed

Condensed from an article by Michael Gehlken

All her life, HSP has compromised Colleen's use of her legs, and a surgery two summers ago that attempted to make her walk stronger with less pain ended up backfiring. With sports, she's found consolation. Colleen, 15, plays on the varsity San Diego Hammer basketball team, which is



run by the San Diego Adaptive Sports Foundation. “I thought ‘What am I going to do?’” said Colleen, a sophomore and straight-A student at High Tech High School. “I have so much energy all the time and so much strength, but I had nowhere to put it. I had no idea what to do. And then once I found sports, it was, ‘Oh, this is perfect! I’ll be OK. It’s not like I lose much. I just lose the ability to walk. I can still play sports. I just can’t walk like I was able to.’” “It drives her crazy not to win,” her coach said. “She’s small, but she’s got unbelievable aggression on the floor. She doesn’t back down from anybody.” Colleen, who lives in Point Loma, CA wants to play tennis and basketball at the University of Arizona, which offers scholarships for adaptive athletes. Sports “is part of my future,” she said. “I couldn’t live without it.”

Off-Road Wheelchairs

On a warm, peaceful October day on the shores of Lake Poinsett in eastern South Dakota, Mike Werner, 56, Sioux Falls pursues his passion, hunting. In 1996, the avid outdoorsman began having trouble with



mobility. He was diagnosed with HSP that has slowly taken his ability to do what he loves. “It’s what I live for,” he says with a smile, “When I’m done working, I want to hunt or fish. I love the outdoors. It’s kind of indescribable. It’s going to be a lifelong memory, a lot of hunting trips are. You forget about the dull times, but you remember the good times.”

The **Action Trackchair** is the brainchild of Tim Swenson of *Action Manufacturing*. His son Jeff was paralyzed in an accident about ten years ago. Like Werner, Jeff was unable to enjoy the great outdoors. So, Tim and his team designed, manufactured and began marketing the innovative machine best described as a wheelchair/ATV hybrid.

Over the years, the concept has become a motto. “Helping the disabled be enabled,” Swenson says with the pride of a father and excitement of businessman.

With the chair, Werner feels a renewed sense of accomplishment. “I’m on fire again! It’s more than

a spark. I don’t know how I’ll find time to work. I just want to get out there and get back the years that I didn’t hunt to my extreme. Now I’m able to do that,” he explains.

The Mobili T Rover consists of a ramp and platform, with a set of rollers embedded in the platform able to use the powered wheels on the owner’s electric wheelchair to drive the front wheels on the Rover. The rear wheels follow up with trailing casters. This setup means the unpowered Rover uses the control and power system from the owner’s wheelchair to do the steering. <http://jalopnik.com/5231519/mobili-t-rover-the-off+road-wheelchair>



New Opportunities for States to Invest in Home and Community-Based Services

Health reform gives states new opportunities to provide home and community-based services (HCBS) through Medicaid, the major payer of long-term services for seniors and people with disabilities. The health reform law, signed by President Obama on March 23, 2010, includes two new Medicaid options that will be available to states beginning in October 2011: The Community First Choice Option and the State Balancing Incentive Payments Program. Both are designed to strengthen non-institutional long-term care services in Medicaid, and both give states added federal matching payments to make it easier for them to expand services and develop new systems to support home and community-based care.

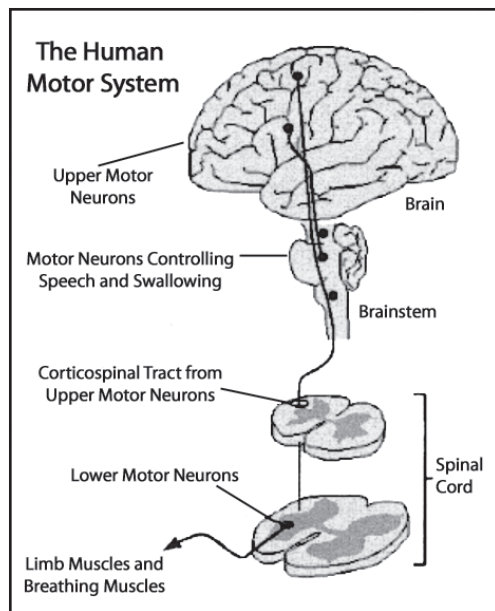
There are several good reasons why states should seriously consider these new opportunities to build home and community-based services capacity in Medicaid. There are state fact sheets which explore the following issues:

- The growing demand and rising costs of long-term care;
- How expansion of home and community-based care can help states; and
- New opportunities that are available in health reform to expand home and community-based services.

Medical Updates

UMN – Understanding My Nerves – Upper Motor Neurons

Ed. note: I asked Dr. Malin Dollinger to write up UMN-101. He graciously submitted the following article so we will all better understand how we're wired.



Hereditary Spastic Paraplegia [HSP] and Primary Lateral Sclerosis [PLS] are upper motor neuron diseases. HSP affects primarily the legs, via the lower spinal cord. PLS may *also* affect the arms (upper spinal cord) and

swallowing and voice (highest spinal cord).

Nerve cells...neurons...control motor function, that make muscles move. The term "nerve cell" means the nerve cell body and the axon, or long nerve fiber that begins in the nerve cell body, on one end, and goes to the next nerve cell body, or to a muscle, on the other end. It's something like a snake, with the head [cell body] on one end, in the brain, and a long body [axon] which makes up the rest of the snake, going down the spinal cord. These axons can be thought of as long thin tubes. They can be very short, an inch or so, or a few feet long, for example the nerves to the legs.

There are two neurons "in a row" that do the job. Upper motor neurons start in the middle of the brain, the "motor cortex," where you decide, for example, to move your right leg forward to walk. An electrical impulse goes down that upper motor neuron, and that neuron ends in the middle of your spinal cord. It there connects with the lower motor neuron, which then goes down to your leg muscle or whatever other muscle you decide to use. A different upper and lower motor neuron "set" is used for every muscle in your body, but in HSP and PLS the problem is in the upper motor neuron, not the lower motor neuron.

The characteristic signs of an upper motor neuron [UMN] problem include increased contraction or "squeezing" of the muscle, called "muscle spasm." This spasm or

spasticity is not coordinated, so the muscles don't contract and relax properly when we try to walk. Another UMN sign is increased reflexes, like when the doctor taps your knees, and your lower leg instantly jumps up, much more than normal. Another sign is "clonus," where the muscles repeatedly contract and relax when stimulated. Another sign is the Babinski sign, where stroking the sole of your foot makes the big toe go up, rather than down as in a normal person. A stroke, or a blood clot in the brain, also produces a Babinski sign, because it is affecting UMN in the brain.

Lower motor neuron disease causes the leg muscles to become "loose" or floppy, not overactive or spastic. Polio is an example. Diseases that cause lower motor neuron problems begin in the spinal cord, where the lower motor neuron starts, rather than in the brain, where the upper motor neuron cell body is located.

ALS involves both upper and lower motor neurons, and is thus different from PLS, which involves only upper motor neurons.

Early ALS resembles PLS, since the symptoms and signs [as described above] are similar. It takes a few years' time to tell them apart.

Misfolding of the SOD1 Protein may Contribute to Sporadic ALS

Robert Brown and Daryl Bosco, neurobiologists and ALS investigators at the University of Massachusetts Medical School in Worcester, coordinated the research team, which published its findings online Oct. 17, 2010, in Nature Neuroscience

A new study suggests misfolding of the SOD1 protein may be a common contributor not only to familial (inherited) amyotrophic lateral sclerosis (ALS) caused by mutations in the SOD1 gene but to the more common sporadic (noninherited) form of the disease as well. If confirmed, the findings could mean that many familial and sporadic forms of ALS share a common factor — protein misfolding. The findings also could mean that therapies being developed to combat mutated SOD1 protein could have application beyond familial SOD1-related ALS.

The team found that SOD1 protein molecules made from a normal (nonmutated) SOD1 gene can assume an abnormal and toxic shape after undergoing a chemical change called oxidation. Once oxidized, they can behave much like SOD1 protein molecules made from mutated SOD1 genes, a known cause of familial ALS. Misfolded SOD1 protein molecules were detected in the spinal cords of four out of nine people who had died of sporadic ALS, and were not seen in 17 people who had died of causes not related to ALS. "We're now working to determine the nature of the

SOD1 modification in patients,” Bosco said. “It may be oxidation and/or other perturbations to the SOD1 protein.”

The scientists performed experiments showing that misfolded SOD1 resulting from either mutated SOD1 genes or SOD1 proteins oxidized in the laboratory can impair the transport of substances along nerve fibers, a problem believed to contribute to the ALS disease process. The investigators on the current study say they have a new antibody, C4F6, which recognizes a different part of the SOD1 molecule from that recognized by previously reported SOD1 antibodies. They say the C4F6 antibody is particularly good at reacting with misfolded SOD1 but not with normally folded SOD1; and that, unlike other methods to detect SOD1 variations, their approach doesn’t artificially alter the SOD1 molecules it targets. However, it isn’t known whether misfolding or aggregation of proteins in the absence of genetic mutations is a fundamental cause of ALS or a downstream effect of a true cause of the disease. The new findings about misfolded SOD1 in sporadic ALS suggest it could at least contribute, if not cause, the disease, since the misfolded SOD1 was found to interfere with transport of substances along nerve fibers.

Meaning for People with ALS

The new findings, if confirmed, point to a common mechanism underlying familial SOD1-related ALS and sporadic ALS, which accounts for 90 percent to 95 percent of all ALS cases. That’s good news for the ALS community, since most laboratory experiments to identify treatments are conducted in rodents with SOD1 mutations, and no one has been certain whether findings in these rodents offer valid predictions of what a treatment will do in humans with the sporadic form of the disease. In addition, misfolded toxic SOD1 provides a clear target against which therapies can be developed.

Drug Approved to Control Emotional Lability

AVANIR Pharmaceuticals Announces FDA Approval of NUEDEXTA™

NUEDEXTA is indicated for the treatment of pseudobulbar affect (PBA). PBA occurs secondary to a variety of otherwise unrelated neurological conditions, and is characterized by involuntary, sudden, and frequent episodes of laughing and/or crying. PBA episodes typically occur out of proportion or incongruent to the patient’s underlying emotional state. “The FDA approval of NUEDEXTA marks an important milestone for people living with PBA, an under-recognized and debilitating neurologic condition,” said Keith Katkin, president and chief executive officer of Avanir. “The approval of NUEDEXTA also marks AVANIR’s transition toward becoming a commercial enterprise, ready to support the successful launch of the first FDA-approved treatment for PBA. We expect that NUEDEXTA will be available by prescription during the first quarter of 2011.”

“PBA is a disabling neurologic condition commonly found in patients with underlying neurologic diseases or injuries. These patients frequently experience embarrassment due to their unpredictable emotional outbursts, leading to disruption of their interpersonal relationships and social isolation,” said Erik P. Piro, MD, PhD, FRCPC, Director of the Section for ALS and Related Disorders at the Cleveland Clinic in Cleveland, Ohio and an investigator in clinical studies evaluating NUEDEXTA. “As a physician who has cared for many patients with PBA, I am pleased that there is now a safe and effective treatment option for PBA that may help these patients regain more control over their daily lives and live with dignity.”

Studies to support the effectiveness of NUEDEXTA were performed in patients with amyotrophic lateral sclerosis (ALS) and multiple sclerosis (MS). NUEDEXTA has not been shown to be safe and effective in other types of emotional lability that can commonly occur, for example, in Alzheimer’s disease and other dementias.

ALS Therapy Development Institute ALS TDI: Open Discussion on ALS Research

By Amy Labbe

ALS Therapy Development Institute (ALS TDI) CEO and Chief Scientific Officer Steve Perrin presented updates on the Institute’s drug development pipeline for ALS during the research symposium portion of a two-day ALS conference.

ALS TDI — pipeline progress Perrin reported that as ALS TDI searches for pharmaceutical partners to help move its lead therapeutic candidate, ALSTDI-00846, into clinical trials, work continues on other drugs of interest. These include: **ALSTDI-000876 (NTF5)**: Evidence suggests this drug is neuroprotective and promotes growth at the neuromuscular junction.

ALSTDI-000866 (apocynin) and **ALSTDI-000896**: Previous studies have produced intriguing results, but neither of these drugs conferred any benefit in testing at ALS TDI.

ALSTDI-000486 and **ALSTDI-00903**: Both drugs aim to modulate the immune system.

‘Hot topics’ in ALS: Fernando Vieira, ALS TDI director of in vivo validation, addressed several areas of ALS research, including: **Stem cells:** Vieira commented on the growing popularity of induced pluripotent stem cells (iPS). He noted that a motor neuron line, created with ALS-affected cells that were regressed to the pluripotent stage and then prompted to develop into neurons, already has been produced for use in drug screening. **Immune system modulation:** Although immune system involvement of neuron support cells called microglia has been implicated by some scientists for years, recent work at ALS TDI has demonstrated a role for other areas of the immune system as well.

Outside experts weigh in: Guest speakers at the symposium included Merit Cudkowicz, an MDA research grantee and director of the MDA/ALS Center at Massachusetts General Hospital in Boston; Gilmore O'Neill, vice president of experimental neurology at Biogen Idec, headquartered in Weston, Mass.; and Clive Svendsen, director of the Cedars-Sinai Regenerative Medicine Institute in Los Angeles. Cudkowicz, who also is a member of MDA's Medical Advisory Committee, said the pace of ALS research is increasing as the discovery of more ALS-associated genes brings in more scientists. She described the ins and outs of clinical trials, including trial design, phases, requirements, oversight, purposes and ALS-specific challenges. Despite the various difficulties associated with testing candidate therapeutics, "Clinical trials are critical to finding the cure for ALS," Cudkowicz said. O'Neill reported on the need to identify and develop ALS clinical biological indicators, called "biomarkers," as a means to deriving more reliable results in clinical trials. The goal in developing therapeutics is to confirm that the drugs are being delivered to the central nervous system, that they are engaging the intended targets, and that they're getting the desired biological and biochemical response, O'Neill said. Ultimately, biomarkers can answer this question for clinicians and researchers: *Is what I think I'm seeing actually the reality of the situation?* Svendsen presented data on an area of study that has "exploded" over the last few years: stem cell therapy in ALS. The first clinical trial to study injection of neural stem cells into the spinal cords of ALS patients, funded by Maryland biotherapeutics company Neuralstem, opened at the MDA/ALS Center at Emory University in Atlanta, in September 2009. Svendsen noted that a second trial already is in the planning stages as a follow-up to the trial at Emory. Preclinical safety studies are expected to begin in the next few months. Patient enrollment is not yet open.

Study Finds Adversity Does Make Us Stronger

Excerpted from an article by Laura Landro

Friedrich Nietzsche was right-sort of. The German philosopher's oft-quoted adage, "What does not destroy me, makes me stronger," was put to the test as part of a national study of the effects of adverse life events on mental health by researchers at the University at Buffalo-the State University of New York and the University of California, Irvine. The study, published in the latest issue of the Journal of Personality and Social Psychology, found that people who had experienced a few adverse events in their lives reported better mental health and well being than people with a history of frequent adversity and people with no history of misfortune. Dr. Seery, a researcher at the Department of Psychology at the University at Buffalo who co-authored the new study says his study shows that, under the right conditions, experiencing some adversity may foster resilience. Age,

personality characteristics and social support systems had no measurable impact on the relationship between adversity and mental health. Adversity can help people develop a "psychological immune system" to help them cope with the slings and arrows that life throws, while those with no experience of adversity may have a hard time dealing with tough times. People who have experienced around two to four adverse events in their lifetimes appeared to be the best off. It is still important to have community and social networks in place to help people deal with the aftermath of adverse events.

Dementia + PLS = New findings

J Neurol Sci. 2010 Aug 30 Clinicopathological characteristics of FTLN-TDP showing corticospinal tract degeneration but lacking lower motor neuron loss.

Kobayashi Z., et al Department of Psychogeriatrics, Tokyo Institute of Psychiatry. Kamikitazawa, Setagaya-ku, Department of Neurology and Neurological Science, Graduate School, Tokyo Medical and Dental University,

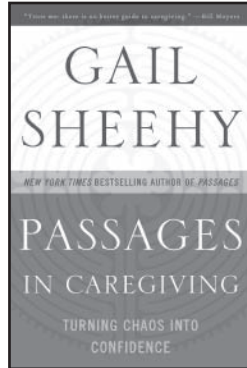
Abstract: The presence of frontotemporal lobar degeneration with TDP-43-positive inclusions (FTLD-TDP) showing corticospinal tract (CST) degeneration but lacking lower motor neuron (LMN) loss has been reported, and the term primary lateral sclerosis (PLS) is used to distinguish motor neuron disease (MND) of these cases from amyotrophic lateral sclerosis (ALS). To date, however, details of clinicopathological findings of FTLN-MND-PLS type (FTLD-MND-P) have not been reported. We evaluated medical records and histopathological findings of ten cases of FTLN-MND-P, in comparison with those of six FTLN-MND-ALS type (FTLD-MND-A) cases. The mean age at onset and disease duration of FTLN-MND-P cases were 54 and 12 years, respectively. The first symptoms were frontotemporal dementia showing behavioral abnormality and/or personality change in five cases, semantic dementia in three cases, progressive non-fluent aphasia in one case, and auditory hallucination in one case. Upper motor neuron signs were clinically identified in six of the ten cases. There were no LMN signs throughout the clinical course in any case. Histopathologically, there was no obvious LMN loss or Bunina bodies in the hypoglossal nucleus or spinal cord in any case, whereas the CST was involved in all cases. The cerebral cortex of the six cases showed type 1 of TDP-43 histology defined by Cairns et al., whereas three cases showed type 3 histology, and one case showed type 2 histology. In all cases, TDP-43 positive neuronal cytoplasmic inclusions were absent or rare in the LMNs, while TDP-43 positive round structures were frequently identified in the neuropil of the spinal cord anterior horn in some cases. This study clarified that FTLN-MND-P cases have characteristic clinicopathological features distinct from those of FTLN-MND-A.

Caring

Book Review of Gail Sheehy's "Passages in Caregiving"

Contributed by Nancy Schaad

Passages in Caregiving: Turning Chaos into Confidence has wonderful information and resources in it. It talks about us all as caregivers and how we will all do it or need it at one time or another. She also is eloquent about palliative care and how it helps people survive much longer than they otherwise would -- it's about getting the help needed to make hard decisions and choose a better quality of life for ourselves. Sheehy wished they would have asked for the help several years before they actually did.



10 Ways To Ask For Help

The Twardowskis

- 1. Brainstorm solutions with friends and family.** Assess the possibilities for help preparing meals - Girl Scout cooking badge; a friend has offered to cook us a complete meal once a week, and we will pay her for the ingredients.
- 2. Build a support network.** Make a list of everyone you know. Talk to friends, neighbors and family members, matching their interest and talents to your needs. If possible, meet with each person individually to discuss how she or he can help.
- 3. Build a support network that also assists your family caregiver.** "We should worry about the caregivers," says Botts. "Caregivers get sick, tired and depressed."
- 4. Tap into service organizations.** I've gotten help from a local Girl Scout troop and teenagers who needed to fulfill service hours for their school.
- 5. Use a care coordination service like myMuscleTeam.** I created a support group e-mail list and send weekly notices telling the "members" what days help is. If I would like to tackle a particular project, I explain what it is and ask for volunteers.

President's Letter (continued from page 2)

It wasn't that long ago that individuals thought of this as science fiction. In fact, Craig Mello at UMass only discovered RNAi in 1998 (and was awarded a Nobel Prize in 2006). Turn the clock back to 2006 and almost no one would be able to describe progress in research as growing human nerve cells with HSP or PLS. Yet today they are.

Ten years ago, there were a handful of genes and mutations for HSP discovered, and they accounted for a small number of HSP cases. As of today, researchers have discovered over 50 genes linked to various forms of HSP, covering over 75-80% of cases.


We should all be proud of the progress we have achieved through our investments since the Foundation was formed. The efforts of the SPF and its members are helping to bring attention to our disorder, and attention brings progress.

Best,

Mike Podanoffsky,
SPF President

- 6. Keep a list of how others can assist you.** I write everything down, from changing a light bulb to picking up a loaf of bread. That way I'm ready if a friend calls me before running an errand. If someone purchases an item for you, pay them when they deliver it.
 - 7. Create short jobs.** In 15 minutes, someone can unload a dishwasher, put the trash out, walk a dog or help me file mail.
 - 8. Copy what other folks are doing.** Attend a local support group. Ask them how they manage.
 - 9. Dial 211 to find community services.** This program is available in 46 states.
 - 10. Think outside the box.** For example, even if you're not a senior citizen, you might be eligible for the services available to them, such as transportation. Perhaps you need a ramp built and don't have the funds to hire a carpenter. Call your local builders association, and ask if they have a community outreach program.
- Be grateful for the help you receive — and express your gratitude.** Send thank-you cards to helpers. Host a dinner (*order take-out*) to recognize the people who make your life easier.

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A graphic for the SPF Annual Conference. It features a red rocket ship with a white star on its side, flying upwards against a blue background filled with white stars. The rocket is labeled 'SPF Research' on its side. A yellow and orange trail follows the rocket's path, with the text 'Rocketing Toward The Cure Through Research' written along it. The SPF logo is prominently displayed in green. Below the logo, the text 'Annual Conference' and 'June 10-12, 2011' is written in black. At the bottom, the location 'Hilton Americas-Houston, Texas' is written in black.

SPF

Annual Conference

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