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SPF 2005 Nat'l Conference and TeamWalk Weekend Columbus, Ohio Sept. 30 - Oct. 2

Sept. 30 – 6:00 pm - Arrival Social
Saturday, Oct. 1 – 8:30 am - National Conference Saturday, Oct. 1 - Evening programs Sunday, Oct. 2 – 10:00 am - TeamWalk Walkathon

Call the Hyatt Regency 1-800-233-1234 to book a room under SP-Foundation for discounted rate.

Contact Paul Brockman fax: 614-895-9987 if you have not received a complete packet in the mail. The packet contains all of the information you need for sponsorships and registration.

The Social and the Conference will be held at the Hyatt Regency. The TeamWalk begins at the Hyatt and proceeds to Goodale Park.

A donation in memory of David Napolin. David enjoyed his print issues of Synapse. The donation was made by friends of David's wife Pat, from Nassau Community College.

Highlights Friday,

Summer 2005

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

We are grateful for two sources of funding assistance for this print issue of Synapse:

or stay later and attend a departure dinner on Saturday. They are holding some handicapped accessible rooms for our group. 405-364-2882. Join us for a great time of fun, food, camaraderie and learning at Andrews Park. We'll gather in one of the pavilions from 10:00 a.m. - 12:00 p.m. Renew friendships, meet others, share stories, learn and have fun! Bring your family and friends, a camera, some cards, games, or snacks if you'd like. After the Connection, we'll walk & roll together on a beautiful sidewalk stroll called Legacy Trail that starts at Andrews Park. Discussions with the local hospital are underway for a luncheon and speaker program after the TeamWalk. Registration for the TeamWalk Connection is only $10 and you will receive a TeamWalk t-shirt.

New York Satellite TeamWalk, Sun., Oct. 16 at Eisenhower Park in East Meadow, Long Island, NY Planned by Betsy Baquet ebaquet@optonline.net 516-520-5906

CA Satellite TeamWalk Saturday, October 8, 2005 Planned by Linda Gentner lkgentner@aol.com 479-651-5676 We have a block of rooms at the Hilton. Eight rooms are Handicap Accessible King, 4 have roll- in showers so make your reservations early. We have an amazing rate of $65.00 which is lower than last year. The Hilton also houses a tennis club and fitness center which is

available to hotel guests. Hilton Pleasanton at The Club Group Name: Spastic Paraplegia Foundation Telephone at the Hotel: 925-463-8000 http://www.hilton.com/en/hi/hotels/index.jhtml;jsessionid=NIEYOD2OEVOLACSGBIV222QKIY FC5UUC?ctyhocn=PLEPHHF TeamWalk will begin at the Valley Community Church, 4455 Del Valley Parkway, Pleasanton, CA

Arrival Dinner -- Friday, October 7 at the

Hilton Saturday activities 10:30am Meet at VCC (church) for coffee,

Satellite TeamWalks across North America!

Canada Satellite TeamWalk, Saturday, Sept. 10 in West Vancouver, BC, Canada Planned by Jean Chambers, 604-990-1060 The event will take place at the Capilano Mobile Home Park. Arrive at 11:00 a.m. to the park's event room to renew friendships and make knew ones, share stories, learn and have fun! Bring your camera, some cards, games, or snacks if you'd like. After socializing a bit, we'll head out for our TeamWalk Walkathon. We'll walk and roll together through the park, wearing out TeamWalk t-shirts, walking and rolling as little or as much as we'd like. After the TeamWalk, we'll enjoy more socializing and unch. A local neurologist has been invited to oin the group to talk about the Baclofen Pump and answer your questions about this spasticity treatment. Afterwards, those who'd ike will go out for dinner at a local restaurant ogether.

New England Satellite TeamWalk Sunday, September 11 Planned by Kathi Geisler kathigeisler1@aol.com 978-256-2673 Rooms: Newport Howard Johnson: http://www.newporthojo.com. Their two-night stay requirement has been waived and our rate for Saturday is $127.50 and for Sunday, $79.20. Call 401-849-2000 and ask for Reservations and request a room from the Team Walk Group: confirmation # P41490. The TeamWalk Connection will be held on the wharf at the Fort Adams State Park. http://www.riparks.com/fortadams.htm >The Newport Mansions are nearby http://www.newportmansions.org/

Oklahoma Satellite TeamWalk, Saturday September 17. Norman OK. Planned by Mark Dvorak mark.dvorak@sp-foundation.org 405-447-6085. For travelers: The local Holiday Inn has set aside a block of rooms for a rate of $89 for those who want to come early for an arrival dinner on Friday

juice, pastries and social kick-off.

11:00am Walk or roll on the sidewalks through quaint downtown Main Street.

Leisurely take in the shops and even a Farmer's Market. From the church through downtown and back to the church is a very easy 2-mile walk (if you want to go the entire

distance). Noon - Lunch back at VCC and more socializing.

Southwest Satellite TeamWalk, Sunday, Oct. 23 in Tempe, AZ at the Kiwanis Park Planned by Bonnie Guzelf bguzelf@cox.net or 480-838-1184. Join us for a great time of fun, food, and camaraderie at the beautiful Kiwanis Park in Tempe AZ. We'll first enjoy a continental breakfast at 10:30 a.m. Renew old friendships, and meet new friends, share stories, and most importantly have fun! Bring your camera, kids, friends and family. Then, we'll walk & roll together around the lake in the park, (7/8 of a mile) walking or rolling as little or as much as you'd like. There will be a picnic lunch in the park after the walk. If you need hotel accommodations, contact me and I will send you information. Hope to see many of you there.

More Upcoming Events!

Aug. 20th 2005, Reelfoot Lake, TN
Planned by Bart Thompson
bthompson2@midsouth.rr.com
http://www.reelfootlake.com/
Accommodations are available at is the Reelfoot Inn, 731-253-6845. They have 24 downstairs units. They have 2 units that are equipped for someone who is wheel chair bound.$59.00 a night; Senior citizens, $55.00, including a Continental Breakfast. Camping at the State Park is another option: 1- 800-250-8617 (first come, first served, no reservations.
http://www.state.tn.us/environment/parks/parks/R
eelfootLake/index.php?activity=Camping

Friday night – we'll meet at Reelfoot Inn and have dinner at one of the local restaurants. Saturday, early breakfast, then a 3 hour cruise on the lake at 9am Free afternoon 6:30 and 7pm we will all meet at the Blue Bank Resort and have dinner.
http://www.bluebankresort.com/search.php
This is one of my favorite places to go. It's where I took my wife for our first real date. Fall in love with your spouse all over again, there is nothing more romantic than holding that special person's hand and watch that big ole sun set over the Lake amongst the cypress trees. It will be a time to forget the pain, sorrow and realization of being handicapped. Just living for that moment.

Autumn in Carolina/ Second Annual SAWCAR Race October 8, in Rural Hall, NC >Planned by Don Wilson don-wilson@earthlink.net 336-969-6748 Rooms:The Holiday Inn Select (Madison Park, Winston-Salem) has reserved a block of 15 rooms, including five accessible (no roll-in showers) rooms in the name of "Autumn in Carolina". The rate for the rooms will be $57.25 plus tax. Ask for rooms from the "Autumn in Carolina" block. The toll free number is 1-800- 553-9595. We will have a hospitality room in the motel on Friday and Saturday evenings. Weekend events: Friday evening - gather in the hospitality room for introductions and conversations, and then the group will share a meal at a nearby restaurant. Saturday program will be in the Fellowship Hall of Kingswood United Methodist Church. One activity will be wheel chair and scooter races under sanction from

SAWCAR (Scooter and Wheel Chair Association of Racing). Racers, start planning your strategy and remember that it takes a lot of expensive technology to maintain those racing machines, so sponsors are needed to help. Everyone will be invited to again share a meal at another nearby restaurant Saturday evening.

also sponsored the coffee bar again this year. When all was said and done---are you ready for this---all expenses paid. Everyone happy. The pearl money and the collections at Spring Fling netted another $2504.00 to the SPF research coffers! How's that for fun and funding?? I thank everyone that sold pearls and wish you could have seen that string. Make no mistake, collecting funds for research is an ongoing activity. Every penny counts. We are in this together and each and everyone of us can lend our help in some way to make this research viable. Check out the fundraising sources on the SPF website. There is always something going on that you can help with. All you newbies are encouraged to take part. Think about attending Spring Fling next year and mark your calendar now. It's always the first weekend in April unless that happens to be Easter and then it is the following weekend. My appreciation to all of you who were at the Fling.

Abilities Expo New York Metro April 15-17

Contributed by Jeanne Young We attended almost a full day Sat. It was a wonderful show, it was my first, I really enjoyed it and highly recommend going if one is close to your area. It was very organized, and I left there feeling more educated,more hopeful,glad that I had PLS if I have to have something. I saw alot of people who were alot worse off than I was, especially the little children born with serious afflictions who never got to have the 50 good years that I did. There were 136 vendors some had every kind of powerchairs you could think of, even more than I've seen online; one of my favorites was a lounge chair with wheels for the beach. They had a lift that can pick you up and put you in and take you out of an inground pool. Christopher and Dana Reeve Paralysis Resource Center, Canine Companions for Independence, Able Newspaper for, by and about the disabled www.ablenews.com were there, as well as van vendors. They also had 37 workshops going over the 3-day period, such as Accessibility Tips for the Home, Aging with a Disability, Differences Between People Born with Physical Disabilities

Abilities Expos Upcoming

http://www.abilitiesexpo.com/IAEBrandManager/ v42/index.cvn

Metro Detroit August 26-28
2005 Novi Expo Center Novi, MI (248) 348-5600

Chicago Metro September 16-18
2005 Donald E Stephens

Convention Center Rosemont, IL
(847) 629-2220

Northern California November 18-20
2005 Santa Clara, CA

Houston, TX December 9-11, 2005
George R. Brown Convention Center,
Houston, TX

Event Reports

Spring Fling, April 1-2 Lovingly organized, run and reported by Ronnie Grove I was pretty lazy at the Fling this year. Annette Lockwood did my job and did it much better than I. Of course, as most of you know by now, if I'm not talking I don't have a clue as to what is being said. I saw a lot of smiles and positive nodding and laughter and a room full of happy people so I know she was saying all the right stuff. We were honored to have Annette, the new SPF president and Mark Weber, the past SPF president.There were 37 in attendance at my best count. We had a good time and learned a lot from each other. We had some items donated by Dorothy Cockrell that brought in $165.00. Annette sold some cards, we ate well, very well, I think; Don and Bettie Jo brought some soft drinks that were well liked; friends at my bank held a 50/50 drawing since I couldn't be there and brought us another $222.00. As it turned out I felt that we had collected enough money to pay for the conference room and expenses without passing the hat. My bank

and Those Who Acquire Them, Mobility Solutions for People with Limited Resources. *Ed. Note: Please check the Upcoming Events section for Expos for the remainder of '05.*

CO Connection, May 21 Contributed by Dale Rutschow Fourteen people showed up at Saint James Church Hall for a great time A big thanks to Dennis Morales for getting us the hall again. This was a show and tell; people were to bring in items or talk about things they do that help them survive. These range from my thoughtful, useful, wonderful subjects to Greg Singer's presentation on "Just because you're a gimp doesn't mean you can't do things that can really hurt". Carl Benham, talked about pain management techniques that he uses. He brought his favorite baseball bat and said that when you get those horrid leg cramps, how just a few good wacks on the head makes you really forget all about them or "going to your happy place" as he put it. But his favorite was licking a live light socket, this had the added benefit of recharging the batteries of his electric w/c! Eric Sansone, who still lift weights, had to agree. He added that just last week, while lifting, got such a bad cramp that he dropped the weights. They landed on his foot and like magic, he forgot all about the cramp, but he didn't know about the ER

being "his happy place". My wife, Jen, talked about how hard it can be to get me out of bed when I'm so stiff and in pain. As a caregiver one must be understanding, gentle, firm but kind and use the devices made to help you. She showed off her favorite transfer device, her cattle prod, and best off it doesn't leave marks! I was a little disturbed by how many wives showed great interest in this... The hit of the day was Greg's presentations. You may have seen his pictures on the list of him water skiing or snow skiing. He brought all his special equipment and also his other hobby, wood carving (which does explain the deep gashes and the missing three fingers). He showed off his skis, they have a seat and platform that he straps his feet to. He even got on, strapped himself in to show how easy it was to balance. We wanted a demo but determined that the baptismal bowl was too small (and we worried what the holy water may do to Greg.) So we got a rope tied it to the back of my van and pulled him down the street. Everything was going great until it dawned on me I had given up driving because of my stiffness and wouldn't you know right then my right leg cramped locking the gas peddle to the floor. I managed a corner at about 70 and Greg was making it getting whipped up the curb, through a flowerbed (looked like a rainbow, like when the sun hits the water rooster tail just right!) but then the rope caught the stop sign... Well Greg wiped out, slid across the street and smacked into a tree. We all ran to see if he was okay, it looked like he was giving a "thumbs up" but it may just be the way his arm was caught in the branch. He was complaining about not being able to move his legs and we said "Well duh, you have HSP" and as far as those few scrapes, remember they say hitting water hard is like hitting concrete and technically blacktop isn't as hard! We got him to his car, gave him a few Advil, used Carl's pain management technique to get him to his "happy place" and told his son not to worry and just get him home. I haven't heard from him so I figure he's fine...

Greater Washington DC Area - SPF Connection June 25. Contributed by Annette Lockwood On Saturday, June 25, the first SPF Connection in the Greater Washington DC Area was held. Eighteen people participated in the meeting. For most of them, it was the first Connection that they attended. Dr. Fishman spoke to the group about Spasticity and Treatment of it. Joy Shewbridge, MMS, PA-C discussed the benefits of an Intrathecal Baclofen Pump and Jackie Sorenson of Medtronic was present to answer any specific questions. Medtronic was kind enough to sponsor the morning coffee service as well. My Physical Therapist was to speak at the meeting but was unable to be there. Thank goodness for Julie Thompson, a retired PT, who offered to speak to the group on Physical Therapy. The speakers did a wonderful job.

Apparently all of us are camera shy because we do not have any pictures to share with

you. Attendees included some familiar names such as Julie, Richard Milbourne, Jeanne and Bill Kroll (who attended along with their close friend Katherine Carroll). Also, Larry, Andrew & Jeanne of the Mercer family joined us along with Chris Tigges, Joan Shoemaker, Janet Cross and her daughter, Susan Klebick. Thanks to everyone for making the first Greater Washington DC Area Connection a success.

CT Connection July 9 Planned and reported by Dolores Carron On Saturday, July 9, 2005 the 20th CT Connection was held in Hartford, CT at the N.E.A.T. (New England Assistive Technology) Marketplace. The 25 attendees included people with PLS, HSP, MD, and MS and their guests as well as 2 Physical Therapists and an RN from an area MDA Clinic. The group was about 50/50- half who'd been with us before and half who met with us for the first time. Judging by the amount of chatter, I think all had an enjoyable time. I know I did. Presentations and tours of the facility, equipment closet (huge), and computer/speaker technology room were given by 4 N.E.A.T. staff members. They'll be having an open house in October with many vendors displaying their assistive equipment and programs. If you are in the area, I'd encourage you to attend it or you can visit them at your convenience. You can get further information at their website www.neatmarketplace.org . They have some very good condition assistive equipment for sale at great prices and will ship to anywhere in the country. They encourage you to 'try it before you buy it' and will make

:hat possible for you to do. The president ınd trainer of Service Dogs International ılso spoke and Takita, the black lab demonstrated some of the tasks that he's ɔeen trained to do as a helper dog. He is still ıot claimed and will be ready for 'adoption' n 3 months. We all fell in love with him!! If ɣou are interested in getting a helper dog, ɣou can contact Keith Mullinar at nullinar@sbcglobal.net .

Armand Legault, CPA and author of "Take It All Off" in a recent MDA Magazine, spoke to us again about various tax and other financial benefits and programs for disabled persons. He is a wonderful resource person and knows about such matters from both sides of the desk-as an insider in the tax world and as a disabled person. Armand has MD, has never walked, and has almost no use of his hands. His van is phenomenal! He speaks 'drive' or 'reverse' and uses a joy stick to drive-forward for drive, back for brake, left and right to steer. His motorized chair can go either in the driver's location or in the passenger's side. He is an inspiration and a really nice person and friend. All in all, it was a great day. Even the monsoon of Friday dissipated and the weather was perfect. My thanks to all who attended for their support and interest.

MEDICAL RESEARCH UPDATES

Research

Snippets

ALS Research Could Lead to Diagnostic Assay Full Story: http://biz.yahoo.com/prnews/050601/new001.htm l?.v=13 Dr. Robert Bowser, director of the ALS Tissue Bank and an Associate Professor of Pathology at the University of Pittsburgh School of Medicine is using a key piece of biomarker discovery equipment known as SELDI to find new ways to identify and eventually treat Amyotrophic Lateral Sclerosis (ALS), a neurodegenerative disease. He is so confident in his ALS research that he recently formed Knopp NeuroSciences to commercially market a more accurate and faster way of diagnosing the fatal disease, which begins by attacking nerve cells in the brain and spine until patients cannot walk, swallow, talk or breathe.

with a different dosage of BTX. The doses were decided on the basis of suggestions in the literature. Outcome measures (Modified Ashworth Scale, Medical Research Council Scale, gait assessment, presence of Achilles tendon clonus, Visual Analogue Scales for Gait Function and Pain, Adverse Effects scale) were applied at baseline, 4 weeks and 4 months after treatment. All the groups showed significant scales scores improvements after treatment with BTX. BTX injections constitute a useful and safe method of improving post-stroke foot spasticity, associated pain, gait speed and function. In particular, the medium BTX dosages (320 UI spread over 2-5 muscles) were found to be both safe and effective in producing long-lasting improvement of spastic foot dysfunction. SOURCE: Neurol Sci. 2005 Apr;26(1):26-31.

Primary lateral sclerosis as a phenotypic manifestation of familial ALS. Brugman F, Wokke JH, Vianney de Jong JM, Franssen H, Faber CG, Van den Berg LH. Department of Neurology, Rudolf Magnus Institute of Neuroscience, University Medical Center Utrecht, GA Utrecht, The Netherlands. f.brugman@neuro.azu.nl http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=15911810 &dopt=Abstract Primary lateral sclerosis (PLS) is a diagnosis of exclusion in patients with progressive spinobulbar spasticity and could be part of the clinical spectrum of ALS. Unlike ALS, which is familial in 5 to 10% of the cases, PLS has been described as a sporadic disorder in adults. The authors report two patients with PLS from unrelated SOD1-negative familial ALS families. These observations provide further evidence that PLS can be linked pathophysiologically to ALS. SOURCE: Neurology. 2005 May 24;64(10):1778-9.

Anti-spasticity Treatments for Children Hagglund G, Andersson S, Duppe H, Pedertsen HL, Nordmark E, Westbom L. Department of Orthopedics, University

Hospital,

Autoimmune Illnesses and Degenerative Diseases

http://www.immed.org/illness/autoimmune_illnes s_research.html Prof. Garth L. Nicolson Rheumatoid Arthritis, Multiple Sclerosis, Amyotrophic Lateral Sclerosis, Inflammatory Bowel Diseases, Scleroderma and other Autoimmune and Degenerative Diseases Autoimmune and Degenerative Diseases are complex, multiorgan diseases of unknown etiology. Although we do not know exactly what causes Autoimmune and Degenerative Diseases, there is increasing evidence that in many patients chronic infections, particularly by certain bacteria and viruses, play an important role in these diseases along with genetic predisposition and immune dysfunction. How could infections be important in Autoimmune Diseases? They could be involved in helping to cause the illness, or they can affect patients by serving as cofactors for the illness (not causing illness on their own but serving as important factors in the disease process) or even as opportunistic infections that increase patient morbidity (sickness) and complications associated with the disease.

Treatment of Spastic Feet with Botox

http://www.ncbi.nlm.nih.gov/entrez/query.fcg ?c md=Retrieve&db=PubMed&list_uids=15877184 &dopt=Abstract A randomised, double-blind, dose-ranging study to evaluate efficacy and safety of three doses of botulinum toxin type A (Botox) for the treatment of spastic foot. Mancini F, Sandrini G, Moglia A, Nappi G, Pacchetti C. Botulinum toxin A (BTX) injections have been used successfully in the treatment of post-stroke foot spasticity, but the optimal dose-response relationship for selected muscles has yet to be established. The aim of this study was to outline beneficial and unwanted effects of three different doses of BTX in the treatment of spastic foot. In this randomised, double-blind, dose-ranging study, 45 spastic feet were randomly allocated to one of three groups, each of which was treated

Lund Department of Orthopedics University Hospital, Malmo and Departments of Physical Therapy Pediatrics, University Hospital, Lund, Sweden. http://www.ncbi.nlm.nih.gov/entrez/query.fcgi ?c md=Retrieve&db=PubMed&list_uids=15931031 &dopt=Abstract Prevention of severe contractures might replace multilevel surgery in cerebral palsy: results of a population-based health care programme and new techniques to reduce spasticity. During the 1990s three new techniques to reduce spasticity and dystonia in children with cerebral palsy (CP) were introduced in southern Sweden: selective dorsal rhizotomy, continuous intrathecal baclofen infusion and botulinum toxin treatment. In 1994 a CP register and a health care programme, aimed to prevent hip dislocation and severe

contractures, were initiated in the area. The total population of children with CP born 1990- 1991, 1992-1993 and 1994-1995 was evaluated and compared at 8 years of age. In non-ambulant children the passive range of motion in hip, knee and ankle improved significantly from the first to the later age groups. Ambulant children had similar range of motion in the three age groups, with almost no severe contractures. The proportion of children treated with orthopaedic surgery for contracture or skeletal torsion deformity decreased from 40 to 15% (P=0.0019). One-fifth of the children with spastic diplegia had been treated with selective dorsal rhizotomy. One-third of the children born 1994-1995 had been treated with botulinum toxin before 8 years of age. With early treatment of spasticity, early non-operative treatment of contracture and prevention of hip dislocation, the need for orthopaedic surgery for contracture or torsion deformity is reduced, and the need for multilevel procedures seems to be eliminated. SOURCE: J Pediatr Orthop B. 2005 Jul;14(4):268-72

Theory on the Possible Cause of Sporadic ALS Acquired nucleic acid changes may trigger sporadic amyotrophic lateral sclerosis. C. Armon.Division of Neurology, Baystate

Medical Center, 759 Chestnut Street, Springfield, Massachusetts 01199, USA. http://www3.interscience.wiley.com/cgi-bin/abstract/110528367/ABSTRACT This article brings together evidence to support the hypothesis that acquired nucleic acid changes are the proximate causes, "triggers," or "initiators" of sporadic amyotrophic lateral sclerosis (ALS). Clinical features that support this hypothesis include focal onset and spread, and the individualized rate of progression. Clues from the epidemiology of sporadic ALS include the increase in its incidence with age, suggesting accrual of time-dependent changes, and the emergence of smoking, a known carcinogen, as its first "more likely than not" exogenous risk factor. The identification of any exogenous risk factor suggests that a large proportion of sporadic cases have a triggering mechanism susceptible to that factor. Ingestion of the products of cycad circinalis has been hypothesized to be implicated in causing Western Pacific ALS. Cycad contains both neurotoxic factors and carcinogens. The dissimilarity of Western Pacific ALS from neurotoxic diseases suggests a greater likelihood that the effects of DNA alkylation are its proximate cause. SOURCE: Muscle Nerve. 2005 Jun 9; [Epub ahead of print]

Neuronal Dysfunction in PLS http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=15829597 &dopt=Abstract *(Note from Mark Weber: The following study was done by Mary Kay Floeter, MD, PhD and her colleagues at NIH. The only reason that this study was able to be done was that at least 25 PLSers stepped up to the plate and volunteered to be subjects in Dr. Floeter's research. Without their help, this study would never have been done. Thanks!!)* Floeter MK, Zhai P, Saigal R, Kim Y, Statland J. EMG section,

National Institute of Neurological Disorders and Stroke, Bethesda, MD, USA. Patients with corticospinal tract dysfunction have slow voluntary movements with brisk stretch

reflexes and spasticity. Previous studies reported reduced firing rates of motor units during voluntary contraction. To assess whether this firing behavior occurs because motor neurons do not respond normally to excitatory inputs, we studied motor units in patients with primary lateral sclerosis, a degenerative syndrome of progressive spasticity. Firing rates were measured from motor units in the wrist extensor muscles at varying levels of voluntary contraction up to 10% maximal force. At each force level, the firing rate was measured with and without added muscle vibration, a maneuver that repetitively activates muscle spindles. In motor units from age-matched control subjects, the firing rate increased with successively stronger contractions as well as with the addition of vibration at each force level. In patients with primary lateral sclerosis, motor unit firing rates remained stable, or in some cases declined, with progressively stronger contractions or with muscle vibration. We conclude that excitatory inputs produce a blunted response in motor neurons in patients with primary lateral sclerosis compared to age-matched controls. The potential explanations include abnormal activation of voltage-activated channels that produce stable membrane plateaus at low voltages, abnormal recruitment of the motor pool, or tonic inhibition of motor neurons.

Primary Lateral Sclerosis, Hereditary Spastic Paraplegia and Amyotrophic Lateral Sclerosis: Discrete Entities or Spectrum? by Michael J. Strong1,* and Paul H. Gordon2 *(Contributed by Jennifer Thomson, as "Fruits from the PLS Symposium")* 1Department of Clinical Neurological Sciences, The University of Western Ontario, and the Cell Biology Research Group, Robarts Research Institute, London, Ontario, Canada; 2Eleanor and Lou Gehrig MDA/ALS Research Center, Department of Neurology, Columbia University, New York, NY, USA. http://www.ingentaconnect.com/content/tandf/als/ 2005/00000006/00000001 Vol 6, Iisue 1 -March 2005. *(ed note: tables and credits have been omitted*

from the excerpts printed below. Please open the article via the link above for all details.) Among the motor neuron diseases, three share the clinical features of prominent upper motor neuron signs - amyotrophic lateral sclerosis (ALS), primary lateral sclerosis (PLS) and the hereditary spastic paraplegias (HSP). While genetic testing can assist in the identification of several variants of the latter, in the remaining cases, including those in which spasticity may be associated with amyotrophy, clinical differentiation of the three disorders may prove difficult. In this paper we review the evidence that these are distinct disorders

and conclude that, for ALS and PLS particularly, there may be justification in considering them as single points along a continuum of multisystem disorders with conspicuous motor neuron involvement. Only through the development and application of exacting clinical diagnostic criteria to epidemiological studies, along with greater numbers of post-mortem examinations, however, will these questions be answered fully. Introduction The motor neuron diseases are a heterogeneous group of disorders that affect either the upper motor neurons (UMN), the lower motor neurons (LMN), or both. There is a divergence of opinion as to whether they represent a continuum of clinically similar disorders sharing a common biological basis, or biologically distinct disorders sharing a limited clinical phenotype. Historically, some have viewed amyotrophic lateral sclerosis (ALS) and primary lateral sclerosis (PLS) as being distinct entities, while others have argued for their interrelatedness. In this article, we review the clinical aspects of these two disorders and the related disorder, hereditary spastic paraplegia (HSP). We will examine the evidence that each may be a unique disease, or conversely, represent but a single point along a continuum of degenerative disease of the motor system. In the purest sense, ALS is a progressive, nearly always fatal disorder that involves both the UMN and LMN, while PLS is held to be a disorder restricted to the UMN in which the clinical course

is sufficiently different from that of ALS to consider it a discrete entity. HSP, distinctive in its clinical manifestations and easily differentiated from both ALS and PLS in most instances, may still present diagnostic challenges when accompanied by amyotrophy or occurring without obvious family history. In each of the disorders, considerable involvement of non-motor systems has been described. Consequently, at least within the category of HSP, there are accepted pure and complicated phenotypes. Such a phenomenological dichotomization provides an opportunity to expand our understanding of these diseases, and to examine whether their pathogeneses may overlap. Primary Lateral Sclerosis The typical clinical features of PLS have been encapsulated by the following: survival is generally longer than that of ALS; bladder dysfunction is rare and usually a late occurrence; the predominant manifestation is purely spinal although a pseudobulbar state may arise; few children are affected; there are no familial late onset cases; the transition to ALS may take over two decades; and overt dementia is exceptional. Because there is currently no defining test, PLS remains a clinical diagnosis. There is an extensive list of disorders to be excluded, which is not too dissimilar from that invoked for either ALS or HSP. The central issue is whether PLS exists as a distinct disease, deserving of a separate nomenclature, or whether an indolent disorder predominantly affecting the UMN represents a very uncommon

manifestation of ALS. As discussed by Rowland, it is truly rare to confirm the diagnosis of PLS, separate from ALS, with a detailed neuropathological examination. The relevance is not merely academic; were the latter hypothesis to be true, the implication is that there is always the possibility that ALS, along with its dire prognostic implications, may appear at some time during the course of PLS. The question of LMN involvement is critical in defining PLS. One caveat is that not all of these

patients may have had a pure disorder of the UMN initially. Nine of the 20 patients described cramps or fasciculations at symptom onset, and six had abnormalities on the initial EMG. Thus, in this cohort of patients, there is evidence of involvement outside of the corticospinal tracts, including LMN signs, albeit less marked than those observed in otherwise typical ALS. Even with LMN findings, however, the overall prognosis was more benign than that of classical ALS. Based on the clinical and neuropathological features of eight cases, Pringle et al. proposed diagnostic criteria for PLS . The authors concluded that the cardinal features of PLS are limited to those associated with degeneration of the descending motor tracts, including spasticity, pseudobulbar affect and spastic dysarthria . Importantly, although higher cortical function was said to be preserved, testing paradigms sensitive to frontotemporal dysfunction were not applied and thus the absence of frontotemporal dysfunction cannot be verified. Among the clinical criteria cited by Pringle et al., the requirement for a disease duration of greater than or equal to 3 years has proved troublesome. Neuropathologically, the time to conversion may also be a moot point because the majority of PLS patients have had the neuropathological features of spinal motor neuron degeneration typical of ALS at autopsy. Recent descriptions of frontotemporal lobar degeneration co-existing with PLS also support the possibility that PLS is but a point in a spectrum of multisystem neurodegenerative diseases. It has not been possible to separate ALS from PLS either clinically or neuropathologically in most instances - a problem compounded by the lack of recent post-mortem examinations on patients with clinical PLS. Hereditary Spastic Paraplegia (HSP) Hereditary spastic paraplegia is a heterogeneous group of disorders sharing a common phenotype of progressive spasticity. A wide range of symptom onset is recognized, spanning infancy to the eighth decade of life. The majority of patients

develop symptoms in the second to fourth decades. Although the prevalence of HSP is not precisely known owing to the indolent nature of the disease, estimates range from 2.0 to 9.6/100,000. The existence of both autosomal recessive (AR) and X-linked variants of HSP may lead to a failure to recognize the familial nature of the spasticity and bring about a misdiagnosis of PLS or a predominantly spastic variant of ALS. Criteria

have been proposed that allow for the diagnosis of HSP in family members at risk for the development of the disease. The obligatory features of HSP include the presence of a family history of a similar disorder, a progressive gait disturbance, lower limb spasticity and extensor plantar responses. In the series described by Harding (1981), common features included lower limb paresis (although paresis greater than spasticity is unusual), mild dorsal column dysfunction, pes cavus malformation, increased upper limb reflexes, mild terminal dysmetria and a loss of ankle deep tendon reflexes. Both distal amyotrophy and paresis of the upper limbs were uncommon clinical features. Harding also separated patients based on age at symptom onset of more or less than 35 years. Those less than age 35 (Type I) had a slowly progressive disease and enjoyed a normal life span, while those with onset after age 35 (Type II) demonstrated a more rapid evolution, often developed abnormal bladder function (50%) and had impairments in sensory function (usually marked by impaired vibratory and joint position sense). Craniobulbar function was spared in both types. In the same series, HSP patients were dichotomized as being either `pure' or `complicated'. In the latter,features of dementia, choreoathetosis, short stature, mental retardation, pigmentary macular degeneration and cataracts could occur. The critical aspect in our understanding of HSP has been the recent explosion in the number of variants of HSP that can now be attributed to specific gene defects. Comprehending how genetic defects bring about a progressive syndrome in which :he major clinical feature is spasticity, and more specifically how HSP can be associated with amyotrophy, will be essential to understanding these disorders. Although an autosomal dominant inheritance pattern would be typical of an HSP, its presence is sufficiently common in ALS that it cannot be reliably used to exclude ALS from the differential. Differentiating HSP from PLS or ALS can be aided by both genetic linkage studies and biochemistry, with the latter allowing for the biochemical categorization of HSP into several large groups sharing common features 24. For example, abnormalities of the formation of myelin and of the embryonic development of the corticospinal tracts seen in X-linked HSP are not among the biological features of ALS. Amyotrophic Lateral Sclerosis (ALS) ALS, the most common motor neuron disease, is age-dependent having a mean onset in the sixth decade. While the average survival from symptom onset in ALS is less than 5 years, this value is misleading in that survival curves are skewed towards short-term survival. Long-term survival may occur, however; some patients have an indolent rate of progression and survival greater than 10 years. In general, longer survival is associated with a younger age at symptom onset (<age 45 years), being male, and having limb onset symptoms in contrast to onset with bulbar dysfunction. There is little regional variability in reported ALS incidence rates, which range between 1.47 and 2.70 per 100,000 individuals (average of 1.89/100,000). After the age of 50, survival curves and incidence curves for males and females converge. Diagnostic criteria have

been created for use in clinical trials in ALS. According to the El Escorial criteria, diagnosis of definite ALS requires evidence of LMN degeneration by clinical, electrophysiological or neuropathological examination; evidence of UMN degeneration by clinical examination; and, progressive spread of symptoms or signs within a region or to other regions, as determined by history or examination in at least three body regions (bulbar, cervical, thoracic or lumbar). These findings must occur in

the absence of electrophysiological, neuroimaging or pathological evidence of other diseases that might explain the signs of LMN and/or UMN degeneration. The diagnostic certainty of ALS can then be based on the extent to which these criteria are fulfilled. There are at least three clinical variants of ALS, including classical sporadic ALS (SALS), a subgroup restricted to the previously endemic focus on the western Pacific island of Guam (ALS-dementia complex of Guam), and familial forms. In keeping with the previous two disorders, ALS is generally considered to be a disorder in which there is a selective loss of motor neuron populations accompanied by degeneration of the corticospinal tracts. Similar to both PLS and HSP, however, impaired cognition like that observed in frontotemporal degeneration is increasingly recognized as a component of ALS.

Conclusions We have described the clinical manifestations of three motor neuron disorders, PLS, HSP and ALS, and in doing so, have attempted to examine whether a significant clinical overlap occurs among the three. If we extend the concept of `pure' HSP to both PLS and ALS, then there may be little in the way of overlap. However, in applying the concept of `complicated' phenotypes, clinical overlap becomes more apparent. Many of the features that define complicated HSP would not be observed in either PLS or ALS, however. Moreover, given our current understanding of the genetics of HSPs, categorization as an HSP is possible for the vast majority of patients with an inherited variant of pure spastic paraplegia. Only in rare autosomal dominant HSP in which distal amyotrophy is prominent in the absence of other features of complicated HSP can the overlap with familial variants of ALS with prolonged survival become an issue. The greater, more controversial, problem lies in separating PLS from ALS. Indeed, there is currently little evidence to support that these are discrete entities, other than disparate prognoses. Modern neuroimaging, neuropsychological testing, neurochemistry and immunohistochemistry indicate that, in most instances, the similarity between these two multisystem disorders outweighs their differences. A cautious explanation would be to view both ALS and PLS as two points in a multisystem degenerative process in which the motor system (either the UMNs or LMNs, or both) can be preferentially involved. PLS, therefore, could simply be an indolent form of ALS. The problem, and the controversy, then

arises in determining which, if any, central feature is the sine qua non of such a spectrum of motor neuron degeneration. Moreover, one then is left to reconcile what may ultimately be a fundamental mismatch between the underlying pathological process and its clinical manifestation. This is highlighted by the overlap of neuropathological features between ALS and PLS, even in those cases in which the Pringle criteria have been adhered to. This issue cannot be answered based solely on the current literature cited above. An approach might be, however, to utilize a clinical categorization of `pure' versus`complicated' disease phenotype. Rigorous, well-designed epidemiologic studies in which every attempt is then made to identify truly pure instances of a progressive UMN degeneration will help determine whether PLS exists in isolation, and define the spectrum of disorders in-between PLS and ALS, along with their different prognoses. An important component of such an undertaking is more dedicated neuropathological study and clinical-anatomic description. The first step in this process is the delineation of consensus diagnostic criteria.

Planning Underway for Initial Stem Cell Safety Trial in ALS ”San Francisco Meeting of Stem Cell Society Highlights State of the Science” Roberta Friedman, PhD, ALSA Research Department Information Coordinator In a preparatory step, ALSA funded researchers have submitted to the U.S. Food and Drug Administration a proposal for a safety study in

ALS patients of a stem cell therapy that is still being tested in animals. A teleconference with the FDA in May clarified what tasks must be performed before that initial human safety study can begin. As detaile by the researchers in the attachment, significant work in large animals must take place successfully before the stem cell strategy can be tested for safety in people with ALS. The safety study will then have to determine that the stem cell treatment is safe, before testing to determine effectiveness in ALS can take place. ALSA is encouraged that planning is underway for stem cell therapy to be tested in ALS. Other equally promising therapeutic avenues are also continuing to be explored in ALSA funded research. On a cautionary note, no published study has yet demonstrated that the stem cell technique proposed for human study has prolonged life or slowed disease progression even in rats modeling the disease. ALSA Science Director and Vice President Lucie Bruijn, Ph.D., said she does not know the time frame for potential safety clinical trials, but ALSA will keep the community informed. Bruijn attended the third annual meeting of The International Society for Stem Cell Research (ISSCR) in San Francisco, dedicated to all aspects of stem cell biology. Leaders in the stem cell field focused discussions on how to nudge stem cells to

become various cell types including neurons, how to grow cells free of contamination that would be appropriate for clinical use, an appreciation of the complexity of creating and caring for these cells in the lab. New technologies and approaches to understanding the biology were discussed. Ian Wilmut, Ph.D., a Scottish researcher at the University of Edinburgh who gained attention by cloning the sheep Dolly, also reported at the meeting on his collaboration with an international group aimed at a better understanding of ALS. Wilmut and his colleagues will take skin cells of people with motor neuron disease, extract the genetic material and place it into donated egg cells, which will be grown for six days. Stem cells from the embryos will be prompted to develop

into nerve cells. Because these cells could incorporate a genetic flaw responsible for the disease, studying the genes might give clues about how the disease developed. The gathering of some 2000 stem cell researchers from around the world also featured a keynote speech by Robert Klein who played a leading role in the development of the California Proposition 71 that has led to that state's $3 billion funding of stem cell investigations independent of the federal government's restrictions on which lines of the cells can be studied with federal funding. Klein emphasized the value and importance of the international society and its meeting in bringing all the world experts together to help move the field forward.

ALSA Techpresenter Contributed by Frank Reyerse For an easy to understand, upbeat summary of the many facets of the work of ALSA, please open and listen to this link. http://onbluestudios.com/alsa/ For those with hard copy, go to your local public library and they should be able to assist you in hearing the content.

LIVING WITH HSP/PLS

A Quote Worth Remembering Shared by Joanne Koenig Coste during her talk in April at the MA-SP Connection "Life is not a journey to the grave with the intention of arriving safely in a pretty and well preserved body, But rather to skid in broadside, thoroughly used up, totally worn out, and loudly proclaiming, 'WOW! What a ride!'" (anon.)

Stretching Exercises Contributed by Dolores Carron Morning Stretching Exercises From Laura Inverarity, “Your Guide to Physical Therapy” Stretching in the morning is a great way to "waken" up your muscles, and get them ready for the day. Stretching loosens up your body while increasing blood flow to your muscles. Incorporating morning stretches into your daily routine is a positive way to begin each day. Stretch #1 1) Sit on the edge of your bed with your feet on the floor. 2) Bend over

reaching your hands toward your feet on the floor. 3) Arch your back. 4) Hold for a count of 10. 5) Repeat 5 more times. Stretch #2 1) Remain seated on the edge of your bed with your feet on the floor. 2) Rotate your neck in a circle touching your ears to your shoulders. 3) Rotate slowly in a clockwise direction 5 times. 4) Rotate slowly in a counter clockwise direction 5 times. Stretch #3 1) Remain seated on the edge of your bed with your feet on the floor. 2) Shrug your shoulders up to your ears. 3) Repeat 10 more times. Stretch #4 1) Stand next to your bed. 2) Lace your fingers together. 3) Raise your hands above your head palms upward. 4) Lift up stretching your rib cage. 5) Hold for a count of 10. 6) Repeat 5 more times. Stretch #5 1) Remain standing next to your bed. 2) Bend over and touch your fingers to your toes while keeping your knees straight. 3) Hold for a count of 10. 4) Repeat 5 more times. Conclusion You are now warmed up and ready to start your day! These exercises can be performed daily to help keep you limber.

Keyboard Solutions

Contributed by
Mata

If any of you are having trouble with repeated characters appearing when you type with a weakened hand, there's a fix under Accessibility Options in Windows XP (and perhaps earlier versions). To get there click Start | Control Panel | Accessibility Options | Accessibility Options (yep, again), then under "Filter Keys" click "Settings." This will get you to a screen where you can choose "Ignore quick keystrokes and slow down the repeat rate" or experiment with other options. Be sure to OK out. I've been doing awkward right-hand-only typing because my lazy left fingers linger too long on the aaaas and ssss, especially. With the change in settings that doesn't happen. However, it won't allow you to quickly backspace or hold down the space bar if you do that a lot. Try it out! Another thing I learned about is "Five Finger Typist" by SoftDawn. It's a software which will teach you to type with one hand on a regular keyboard without having to look at the keyboard. I see it for $70 on the Web, but there's a free trial version at http://members.ozemail.com.au/~softdawn/s oftwa re/windload.html.

Baclofen Pump- and announcing Miss February Contributed by Kim Richman I've had my pump since July of 2004 so I'm really a newbie but it's something I'm glad I did. The pump does not help me at all with one of my primary issues- that of pain. However, the pump has done a tremendous amount to help me in another area- in simple functionality- my gait- without it I don't know that I'd be walking now- honestly. I'm very thankful for technology and research and glad that I had the surgery. I recently entered a contest that a website was sponsoring. WWW.ExploringSpasticity.com was offering a contest where you could write an essay about the courage it takes to deal with the changes wrought by spasticity. I decided to give it a shot. Well, Everyone, it's not the Sports Illustrated Swimsuit Edition, but at age 39,I've

become a February Calendar Girl! (LOL) Look out for next year's contest- and consider entering.

National Council on Aging Call your nearest **regional office** or click on https://ssl3.benefitscheckup.org/ to explore benefits to which you are entitled.

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Great Nutritional Drink
Contributed by Cheryl (HSP in Maine)

I would like to share information so that you can enjoy this tasty AND healthy milkshake...As was previously recommended to me, I purchased a nutritional drink mix called GenSoy at my local grocery store. It comes in French Vanilla (kind I prefer), Chocolate, and Plain and can be mixed with milk (my preference is skim milk), juice, or water, along with berries and/or ice cubes--all can be blended and crushed in a blender. This drink has no fat and provides protein and many essential vitamins and minerals. My preference is to mix French Vanilla with raspberries or strawberries.

FUNDRAISING

1,000,000 Pennies for Research

Contributed by Ronnie Grove frogrove@pennswoods.net At the beginning of May I pitched an idea for a new fund raiser. It is called Saving Pennies For SPF. The goal is for all of us to help collect one million pennies by Spring Fing 2006. That's $10,00.00 and here's how it will work. May was our warm-up month and warm up we did! We collected one hundred sixty four thousand four hundred eighty nine pennies ($1644.89). Kick off will officially begin June 1st. I would like to have ten teams (I have nine) with at least ten members to a team for a break down that is manageable. Each team would need to reach a goal of $1,000.00 dollars over the next ten months. A mere $100.00 a month and only $10.00 per team member or even less if a team recruits more than ten members. Since May was only a trial run and we got off to a wonderful start I think we can do this easily. However, we will need all the help we can get. If you haven't signed up for a team please do so as soon as possible. If you would like to be a

captain let me know. Captains on board are Jean Chambers, Sue Me Melchor, Eva Wright, Lili Plaez, Sarah Duncan, Charlotte Hubbard, Jeannie Young, SusieQ, and Ronnie Grove. >There are ways to help other than collecting pennies. You might want to just pledge an amount each month or ask friends and family to pledge a certain amount monthly or just ask for a one time check anytime between now and April 2006. You might ask for a check for your birthday, Christmas or any other gift giving occasion. If you collect pennies please report your total to your captain monthly so we can keep track of our goal. When pennies are cashed in you may send your check either to your captain or to the SPF Treasurer- David Lewis, PO Box 1208, Fortson, GA 31808. Be sure to mark your check Penny Drive or Penny Campaign. We all win when we can walk again.

Hand Crafted Quilt Made for SPF Raffle

This beautiful Turning Leaves quilt was made by Northern Ontario friends of Theresa Foley. Theresa lives in Northern Ontario, Canada, and has HSP. Four other of her family members also have HSP. Theresa had belonged to the Quilting Club about 5 years ago before being diagnosed with HSP. She approached the president to see if they would make a quilt for the SP Foundation to raffle or to auction for research. It was nearly a year before it was approved and Theresa paid for

the material and the quilting ladies have just finished it. The quilt was presented to Theresa as a gift to SP Foundation to raffle or auction. It is called Turning Leaves and it is 6’ by 7’4” (large double). As you can see by the colors (colours in Canada) it will go

nicely in any bedroom. Wouldn’t it make a wonderful holiday gift? We are setting up a raffle and will pull the winning ticket after all the TeamWalks are completed. Our coordinators are Board Members Linda Gentner (USA) and Jean Chambers (Canada). Please send a check made out to SPF to the appropriate address listed below. The raffle tickets may be printed by clicking below the ticket image then printing one or more pages of the enlarged image. However, you will need to constrain the print image size to 7.7" wide by 10" high before printing.
Tickets are: 1 for $5; 5 for $20; 10 for $30 Checks should be made payable to SPF and sent to: Linda Gentner 1605 Goularte Place Fremont, CA 94539
Jean Chambers 226 Hiawatha Drive West Vancouver, B.C. Canada V7P 1E1