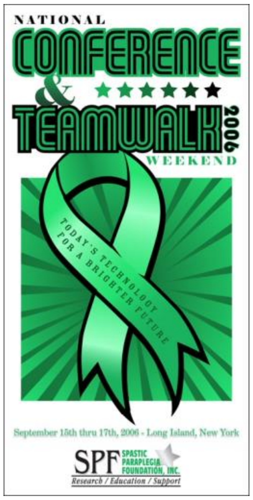
Synapse

Summer 2006

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

SPF 2006 National Conference & TeamWalk Weekend
Long Island, NY; September 15-17



Location & Accommodations:
The Marriott Hotel in Melville, NY (Long Island). Hotel rooms are $99 per night. Call 1-800-228-9290 and ask for group code SPASPAA for this discounted rate. Both JFK and LaGuardia airports are about the same distance from Melville. There is no hotel shuttle service. Help will be made available for individuals needing transportation assistance.

Betsy Baquet is Chair of the weekend. Please call her with questions: 516-520-5906.

Schedule:

September 15th 7:00 p.m. Arrival Dinner in a hotel function room. $25 per person.
(A vegetarian option will be available.)

September 16th 8:30 a.m. - 4:30 p.m.
National Conference
"Today's Technology for a Brighter Future"

Speakers for the day are:
Research, Clinical Information and Treatment

- SPF Medical Advisor and Research Award Recipient John K. Fink, M.D.
- SPF Research Award Recipient Brett Peter Lauring, M.D., Ph.D.
- SPF Medical Advisory Board member Mark Gudesblatt, M.D.

Physical Rehab. And Assistive Devices:

- Craig Rosenberg, M.D.
- James Megna, P.T.

Break-Out Sessions for Patients and Caregivers:

- Deborah Benson, Ph.D.
- Jean Elbaum, Ph.D.

The Conference fee is $50 per person, which includes the conference and materials, continental breakfast and lunch. Saturday Dinner outing to a local restaurant with others if you wish

September 17th 10:00 a.m.TeamWalk For Our Cures Wantagh Park

Registration information for both the National Conference and any TeamWalk for those with computers may be found at http://www.sp-foundation.org/ For those who receive a hard copy of Synapse, a registration form will be mailed to them.

Please be sure to be a part of the "Add 6 in 06" effort to find new sponsors. The sponsorship form is with the TeamWalk registration form for those receiving hard copy.

SP FOUNDATION

A Letter from our SP President

So far this year, we have added 26 new community members to our list, a warm welcome to all. We continue to grow as more people become aware of our organization. Through June 2006, we have raised over $100,000, the breakdown by major initiative is:

$ 25,500 – Year End Appeal Letter
$ 18,100 - Memorials
$ 15,200 - Annual Report/Membership Drive
$ 11,100 – 2006 TeamWalk
$ 10,000 – RGM Golf Classic Memorial
$ 9,100 – Pennies
$ 8,070 – ExxonMobil Workplace Giving

It is sad to see that Memorials are so high on the list. I want to extend our sincere condolences as well as thanks to the families of Charles Grove (Ronnie's Father), Gerald Leary, Hazel Lewis (David's mother), Katherine Parker, Kathleen Richardson and William Swain.

As you can see, we have already received monies for the 2006 TeamWalk. The Committee and Local TeamWalk Coordinators are busy putting together plans for the National Conference and TeamWalks. Soon you will be receiving the brochures in the mail for the National Conference/TeamWalk Weekend and the local TeamWalks. For a truly successful TeamWalk, everyone needs to participate and obtain sponsorships. You can attend the National Conference/TeamWalk in Long Island in September or one of the 10 Local TeamWalk Connections. If you are unable to attend, please join the team as a Walker by Proxy. If you have any questions or suggestions for TeamWalk, please send a note to Teamwalk@sp-foundation.org or call 703-495-9261. Please register online or use the registration forms that you will receive in the mail in the near future.

As mentioned in the previous Synapse, SPF is searching for a professional fundraiser as an Executive Director. We are hoping to start screening candidates in August. Once in place, the Executive Director will help coordinate, plan and launch new and existing fundraising events.

Carolyn Sartain-Anderson is holding a Benefit Dinner on August 12 in Southampton, NY to Celebrate turning 60 and to Benefit SPF. Carolyn has invited high school classmates and friends to the dinner. Our wishes go out to Carolyn and her friends for a very successful and exciting evening in August.

Annette Lockwood

Pocket Change for PLS/HSP

Contributed by Sue Meholick

We are continuing the penny campaign that Ronnie came up with and we raised over $16,000 with last year. We are doing regions this time--so if you would like to be a captain, let me know. Here are the captains so far:

Northeast - Sue Meholick (814)653-8566; 256 Powers Street, Reynoldsville, PA 15851; momofboysonly@yahoo.com
Southwest – Shell Fischer (505)885-1289; MUDLUPII@hotmail.com
Midwest – Jackie Wellman; hoppywell@aol.com

EVENTS

Event Reports

Burlington, MA, May 21

Leigh Hochberg, MD, PHD, talked about the BrainGate clinical trials to test a brain computer interface that translates thoughts into making things happen - to control devices using your thoughts! Currently, the trial underway is for ALS patients. "The goals of this study are to investigate the safety and feasibility of the BrainGate(TM) System in people living with ALS or other motor neuron diseases. If study participants are able to gain control over a computer cursor with the BrainGate System, this could open the door to controlling a broad range of other devices, including assistive communication devices, speech synthesizers, and, eventually, devices to re-enable limb movement. Furthermore, by recording neural activity over time in participants with ALS, this study may help to provide additional insights into the neurodegenerative process that occurs with this complex disease." The audience of about 30 was fascinated to learn about the possibilities this technology might open.

Regional TeamWalks & Other Upcoming SP Events

Note: All TeamWalks have a $10 per person registration fee.

August 12, Saturday **- Southampton, Long Island** Southampton High Class of 1964 60th Birthday Bash/Benefit for the Spastic Paraplegia Foundation(SPF) - 6 p.m.- ? Cocktail Benefit held at my family home in Southampton, Long Island (65 Lewis Street).
We are asking for donations, rather than an admission fee.
Contact Carolyn Sartain Anderson - 772-388-8499
beachmusic1012@bellsouth.net

August 20 – Lexington, MA TeamWalk
Kathi Geisler at kathigeisler1@aol.com and 978-256-2673
Gather at the Lexington Battle Green for the 11 a.m. walk.

August 26 – Nashville, TN TeamWalk
Jim Sheorn jmsheorn@comcast.net and 615-479-7369
Centennial Park

Sep.8-9 - Toledo, OH TeamWalk
Moira Franchetti
_moirafranchetti@sbcglobal.net and (419) 865-0517
Wildwood Metropark at 10:00 a.m. A shelter house has been reserved. A Friday night dinner is being planned.

Sep. 8-9 - Thomasville, GA SP Connection and TeamWalk
Jane Anne King _gking@rose.net and 229- 227-0558 A Friday Arrival Dinner at Ryan's Steak House at 6:30 p.m. Individuals will pay for their own dinners there but registration is needed to reserve enough seats.
1:30 Luncheon at the Plaza Restaurant, $10 paid in advance.
TeamWalk at theYMCA after lunch.
There is a football game that day! If you need a hotel room, book immediately.

Hotels in the area with accessible rooms are the Hampton Inn (229-227-0040), Comfort Inn (229-228-5555), Thomasville Inn (229-227-1111) and the Quality Inn Suite (229-225-2134).

Sept. 9 Annapolis MD TeamWalk
Contact Shannon Gregory at shannon.e.gregory@hq02.usace.army.mil The TeamWalk will be held at Quiet Waters Park. There will be an Arrival and Registration Period from 12:00 - 1:00 with the TeamWalk beginning afterwards. A Celebration Picnic will be at 3:00 and include refreshments and a special program.

Sept. 15-17 Wataugh, LI, NY SP National Conference and TeamWalk
Please see separate article.

Sept.15-16 -Norman, OK TeamWalk and Weekend Gathering
Mark Dvorak - czechmarkmhd@yahoo.com , 405-447-6085
The local Holiday Inn has set aside a block of rooms, some handicapped accessible, for a rate of $89. The rate of $89 applies “ONLY” if 5 or more rooms are reserved. Call 405-364-2882 by August 31st to reserve your room in this block. Be sure to mention that you are coming to Norman for the Spastic Paraplegia Foundation’s TeamWalk
A Friday Arrival Dinner, and a Saturday Night Dinner are being planned to take place just down the street from the hotel. The TeamWalk is to be held Saturday, September the 16th. We will start gathering at the park shelter at 10:00 am and we will start “Walking and Rolling” on the Legacy Trail at 11:00 am.

Sept. 15-16 - Pleasanton, CA TeamWalk and Run
Linda Gentner lkgentner@aol.com and 510-651-5676
Friday Night Arrival Dinner at the nearby Hilton Pleasanton Hotel. Rooms are set aside for our group at a rate of just $65 until September 8 (Spastic Paraplegia Foundation group). The Hilton has ten handicapped accessible rooms and two of them have roll-in showers. Their phone number is 925-463-8000 or call the Hilton worldwide services at 800-445-8667.
The TeamWalk Connection will be held at the Valley Community Church in Pleasanton. As in prior years, Registration and Meet & Greet will include a continental breakfast at the Valley Community Church at 10:30 a.m., followed by a TeamWalk down Main Street. Walkers will meet back at the Church for lunch. We are very excited that this year that our TeamWalk be held in conjunction with a Run event organized by church, with proceeds from it to support SPF!

5th Autumn in Carolina/ 3rd Annual SAWCAR Race October 6-7, 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 $64.00 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.

Oct. 13-15 - Tampa, FL Women's Retreat and TeamWalk
Kathi Geisler kathigeisler1@aol.com and 978-256-2673
We've reserved rooms at the Best Western Hotel, $75 each. The hotel provides shuttle service. (http://book.bestwestern.com/bestwestern/productInfo.do?propertyCode=10325#null)
An Arrival Dinner will be on Friday evening. Registration price to attend the Saturday retreat is (check made out to SPF). $55. Tawny (has PLS) is a former clinician and is taking the lead on the Saturday retreat agenda.
The TeamWalk will begin as a Breakfast Connection at 10:00 at the restaurant at the Best Western Westshore Hotel in Tampa. TeamWalk will then be held at Friendship Trailbridge which overlooks Tampa Bay.

Nov.3-4 Raleigh, NC TeamWalk & Run
Sarah Witt _srwitt@yahoo.com 919-848-0582
A Friday Arrival Dinner is being planned.
The TeamWalk will be at 10:00 a.m. at the centennial campus of NC State University at 10:00. We are excited that it will be in partnership with the NC Roadrunner's Club, with those proceeds also supporting SPF Research Awards! The event will feature a 1-mile competitive race for athletic runners with the Roadrunners Club; a 1-mile (or however far you want to go) TeamWalk for our community members to walk roll or stroll; and a kid's race. Our reserved area includes a covered plaza, open field, and a wide boulevard**.** We are honored that Dr. Bedlack from Duke University, a researcher on motor neuron diseases, will be attending the TeamWalk, as well as a physical therapist who specializes in spasticity management. They will share the latest in areas of research and treatment.

Date TBD Eugene, OR
Contact Karen Long at LaRue2034@aol.com 541-689-9643.

LIVING WITH HSP/PLS AND CAREGIVING

SP Dinner with the Reyerse Family

On June 19, Annette Lockwood, SP President and Jim and Thurza Campbell had the great pleasure of meeting Frank Reyerse's family in Washington, DC. It was wonderful to hear stories about Frank from Joanne, Frank's wife, and 9 other family members. Frank would spend 10 hours a day working on his data base! Jim Campbell was particularly happy to meet Joanne, since they have corresponded via email so much as Jim took over management of the site. (Photos on page 16.)
If you are not in the data base, please go to http://www.geocities.com/freyerse/index.html and send your information today.
Here's what you do:
1. Enter the site above.
2. On the Home page, select Click Here
3. That should open a window entitled: PLS/HSP/ALS Data Base & Guest Book
4. Fill in the information requested by the form, then click Send.

That will automatically send an email to *SynapsePLS@comcast.net* that will contain all of the form's information that we can then use to update your entry in the worldwide data base.

PLSer's Story

Contributed by Linda Gentner

You need to read about Charlie & June Gray. She always signs her posts "June Bug" -- quite a couple! Want to meet them in person? You can, because along with Jane Anne King, they are helping coordinate the Thomasville, GA TeamWalk on September 9th.

"Because He Will Rise Again, Charlie Plays On"

Excerpted from an article by By David Adlerstein, Times City Editor

He played on Chipola Junior College's last football team in 1951-52, and then when he was drafted during the Korean War quarterbacked a team full of All-Americans and future pro players for the champion Brigade Rams at Fort Benning. He's been a loving father to two daughters and commanded a football squad of 10 young men who fought for every yard, and for the pride of this nation. But these days, Charlie Gray's opponent doesn't wear shoulder pads or fire a rifle, and calls itself by the mysterious initials, PLS.

This tough opponent entered Charlie's life nearly five years ago. It's cracked his pelvis and put him in a wheelchair, forced him to use a walker and sent his legs into spasms, and made his voice difficult to understand. But like the Special Troops Command team in 1954 that fell 41-0 after Charlie threw four TDs, including a 44-yard strike to Ed Hamilton and a 44-yard missile to John Middleton, this PLS is getting its butt kicked all over the field, one day at a time, day after day. "I'm okay," said Charlie, after hanging out with his daughter, Pam Nobles, where she works at Apalachicola Physical Therapy. "I will walk and run again. This is my slogan, 'Because I will rise again.'"

Three days a week, Charlie welcomes a physical therapist into the Grays' Tallahassee home to help him tackle those exercises that can master Primary Lateral Sclerosis, a motor neuron-related disease so rare that only one person in 10 million is newly-diagnosed each year.

"She showed him how to walk," said Nobles. "She 'kills' him but that's what they're supposed to do, make you work." In the games, when it was hard, he'd say 'Suck it up,'" Charlie said. "I'm sucking it up. When my physical therapist makes me do things, it's hard."

As tough as PLS is for Charlie, it's a challenge for June, who has stood right alongside her husband the whole nine yards. At 6'3" tall, and a playing weight of 178 pounds, Charlie has the athleticism that helps him confront PLS, but with progressive deterioration of the motor neurons, it has its moments. "You have to have a lot of humor with this disease," said June, recalling the time her husband had trouble once he got into the hot tub outside with the help of a lift, and the emergency squad was summoned.

"That was an experience," said June. "The firemen dropped their britches and got in the pool. All the neighbors came over to see this. They said we should charge admission."

"We just take each day as it comes," she said. "The days are never the same. Every day's a different day." Her husband makes sure she knows how much he cherishes her steady hand.

"She's been so good," said Charlie. "I've had a wonderful life, beyond any dream I've ever had. It's been a good run. And it's been hard on June." June in turn knows sometimes she has to exercise tough love on her husband, so that he can make the most of living with PLS."They're going to turn me into the humane society if I don't quit being mean," she joked.

The Relaxation Response

Ed. Note: In June I was fortunate to hear Dr. Ann Webster of The Mind Body Institute and Harvard Med. School speak at our monthly ALS Support Group. Below I have copied some of The Mind Body Institute's technique for you. Even though the web site does not mention *neurological diseases as ones they treat, their stress management process can be very helpful.*

The Mind Body Institute
http://www.mbmi.org/home/ 617-991-0102

Elicitation of the relaxation response is actually quite easy. There are two essential steps:

1.Repetition of a word, sound, phrase, prayer, or muscular activity.
2.Passive disregard of everyday thoughts that inevitably come to mind and the return to your repetition.

The following is the generic technique taught at the Mind/Body Medical Institute:

1.Pick a focus word, short phrase, or prayer that is firmly rooted in your belief system, such as "one," "peace," "The Lord is my shepherd," "Hail Mary full of grace," or "shalom."
2. Sit quietly in a comfortable position.
3. Close your eyes.
4.Relax your muscles, progressing from your feet to your calves, thighs, abdomen, shoulders, head, and neck.
5.Breathe slowly and naturally, and as you do, say your focus word, sound, phrase, or prayer silently to yourself as you exhale.
6.Assume a passive attitude. Don't worry about how well you're doing. When other thoughts come to mind, simply say to yourself, "Oh well," and gently return to your repetition.
7.Continue for ten to 20 minutes.
8.Do not stand immediately. Continue sitting quietly for a minute or so, allowing other thoughts to return. Then open your eyes and sit for another minute before rising.
9.Practice the technique once or twice daily. Good times to do so are before breakfast and before dinner.

Regular elicitation of the relaxation response has been scientifically proven to be an effective treatment for a wide range of stress-related disorders. In fact, to the extent that any disease is caused or made worse by stress, the relaxation response can help.

Other techniques for evoking the relaxation response are: Imagery, Progressive muscle relaxation, Repetitive prayer, Mindfulness meditation, Repetitive physical exercises, Breath focus.

J –WALKER, A Baby Sitter Aid

Contributed by Flora Brand

I'm going to try to take care of my new grandson and had to think of a way that I could transport him from one room to another. So Doug bought a brand new walker at a garage sale for $20 and converted it to the "J -WALKER" J standing for my grandson Jack.

I never say "never" just have to figure out a new way of handling things differently. (Photo on page 16.)

Links and Phone Numbers that Might be Useful

Center for Health Care Strategies - Improving the quality of publicly financed care. They have a great resource library and information on health plans, community based organizations and state agencies! Visit www.chcs.org or call 609-895-8101.

Office of Disability Employment Policy - Independent federal agency that acts as a national source of information and assistance regarding job listings, workers rights, advocacy, employment services and various programs and initiatives. Visit www.dol.gov. or call 1-866-4-USA-DOL.

Health Insurance Association of America - National trade organization representing nearly 1300 member companies providing health benefits to millions of Americans. Many helpful links and resources. Visit www.hiaa.org or call 800-509-4422.

Life in Motion Letter Writing Campaign – SPF needs your support

Contributed by Annette Lockwood

To commemorate Movement Disorders Awareness Month this October, the Life in Motion campaign is sponsoring the first-ever Movement Disorders Patient Summit in Washington, D.C. on September 14th and 15th. The purpose is to help raise awareness about movement disorders among members of Congress and the media. Representatives from the Life in Motion Coalition, as well as patients, advocates, physicians and other healthcare professionals will be attending the Summit to learn about the latest advancements in treatment options, diagnosis and other quality of life challenges and coping strategies. Congressman Danny Davis (D-IL) will be the keynote speaker at the Patient Summit. I am planning to attend as much of this forum as possible before I leave for our National Conference and TeamWalk in Long Island. In an effort to raise awareness and encourage participation in this forum, SPF is participating in the Life in Motion Congressional letter-writing campaign. How can you help? Phone toll free 866-546-3136 or click on: http://www.life-inmotion.org/lwc/lwc.asp where a form appears, followed by a letter to your Congressional Rep. Please be sure to check off SPF when asked what organization you are supporting. SPF will be awarded funds from the Life in Motion campaign based on the number of individual letters our organization generates. This is an easy way to raise awareness of movement disorders and help SPF at the same time. It took me less than 5 minutes to add my information, read the letter and submit it for processing.

You might also check out http://www.wemove.org/ for comprehensive information on movement disorders.

PT for PLS

Contributed by: Dolores Carron

It is important that you find a PT who specializes in neuro disorders and is very familiar with treating spasticity. Some PT exercises and maneuvers can make our symptoms worse. Our needs are very different than for someone who's recovering from a stroke, or a broken bone, etc. You can go to the American Physical Therapy Association web-site http://www.apta.org/AM/Template.cfm?Section=Find_a_PT&Template=/APTAAPPS/FindAPT/findaptsearch.cfm and click on "choosing a PT" on the left side of the screen. You'll get another page and in item #2, there is a place to "click here" to find a PT in your area. It prompts you to choose the expertise, so choose "neurologic". Then you enter your zip code and the number of miles that you're willing to travel and you'll get a list of providers who offer the treatment you're seeking. 1-800-999-2782.

PT Stretching Exercises

Contributed by Kathi Geisler

Do these routines 3 times a day:

1. For inside thigh stretching: Sit on the floor with one leg stretched straight out in front of you and the other leg bent in so that the bottom of its foot touches the knee of the stretched out leg. Push gently down on that knee so that that leg's inner thigh is stretched. Hold 45 seconds. Do 3 reps.
2. Butterfly Stretch, also for inner thigh stretch: Sit with knees out to the side and bottoms of feet pulled in and touching. Use hands to push gently down on knees and stretch thigh. Hold 45 seconds. 3 reps.
3. Bridge Butt lift: Lie down with your legs bent up. Tighten the buttocks and lift them up in the air so your body is slanted and straight. Hold a few seconds. Do 10 of these. 2 sets.

Chronic vs Curable Disease

By Richard M. Cohen

Clean at last. The third colonoscopy was a charm. Two bouts of colon cancer have ended, the curtain fallen. The houselights are up. Nobody has noticed that I am still on stage. A chronic illness remains. Multiple Sclerosis, my longtime companion, has resumed its lowly position in the hierarchy of suffering.Chronic illness is driven from the stage by the acute threat. Its plotline is tedious because action is slow and the story rarely varies. Attention spans are short, and the drama can take years to play out. When recovery from a life-threatening illness comes, that tired old standby remains. Turning tragedy into comedy is one option for coping. Creeping, crawling illness takes me to the theater of the absurd. Belly laughs sustain me.

Orphan afflictions become the long haul. They have little cachet but afflict the many. These diseases are boring, not the stuff of movies. The Big C is a proven box office winner. Remember the last hot big-budget film about a man with crippling arthritis or a woman with excruciating shingles? I don't.And a public does not understand or appreciate the pace or pain of slow sickness. Many diseases compromise the ability to eat and digest, to walk and to speak and a host of other functions. These conditions remain private because most of us tire of talking, and no one can see the truth of another person's life.

We are left to battle insurance companies that resist the steady costs of endless care and the employers who quickly tire of our bad days. We are compromised. We become a hidden population. We are invisible, except to our bosses and colleagues and others we engage. I have trouble walking. Don can barely eat. Susan has memory problems. We will live another day, but the routines that others take for granted will challenge and occasionally conquer us. We can only acknowledge our difficult journeys to ourselves in a whisper and move forward with humor and grace.

Financial Assistance for New Vehicle Modification

The grueling experience of buying a new car is enough to make almost any wallet tremble. Everything is extra. Now add on the costs of making that vehicle accessible, and suddenly that trembling wallet goes into cardiac arrest. Fortunately many vehicle manufacturers have special programs to help offset some of the costs associated with modifying a purchased (or in some cases leased) new vehicle to make it wheelchair accessible. Leased vehicles must have the lessor's written approval prior to adapting the vehicle.

GM Mobility Reimbursement Program

(800) 323-9935 / *www.gm.com*

General Motors may provide up to $1,000 for adaptive modification through its GM Mobility Reimbursement Program. If an individual purchases or leases a new and unused (not previously titled) 2005, 2006, or 2007 GM car, SUV, van, or truck through an authorized dealer by September 30, 2006.

Ford Mobility Motoring Program

(800) 952-2248 / *www.mobilitymotoringprogram.com*

Ford Motor Company created the Ford Mobility Motoring Program which offers up to $1,000 in adaptive assistance and has a customized assessment center with an adaptive equipment dealer and installer listing.

Chrysler Automobility Program

(800) 255-9877 / *www.chrysler.com*

Daimler Chrysler’s Automobility Program also offers up to $1,000 to help with costs of installing adaptive equipment on a purchased or leased 2004, 2005 or 2006 new and eligible Chrysler, Jeep and Dodge vehicles.

New NIH Testing Protocol for Cognitive Changes in PLS has Begun

Contributed by Thurza Campbell

Study title: Cortical Function in Primary Lateral Sclerosis and ALS #06-N-0174

Dr. Mary K. Floeter is in charge of this protocol. Contact her at floeterm@ninds.nih.gov or call 301-496-7428 to inquire. During the course of two days, I took the following tests: MRI – 45 minutes; EEG (HMCS or MRCP?) – 2 ½ hours; Neurological Exam – 2 hours; Cognitive Psych testing - 3 ½ hours – standardized forms and questions; Psychiatric Evaluation 1 ½ hours; Final wrap up – 15 minutes. NIH pays airfare, lodging and $20 per person per day for meals for you and a companion. They also have a free shuttle from Dulles Airport to the motel and NIH. (Photos on page 16.)

MEDICAL UPDATES

Internet Handbook of Neurology

This is a comprehensive list of diseases and procedures.

http://www.neuropat.dote.hu/nmd.htm

A Letter from the Primary Lateral Sclerosis Nurse at NWU

Dear PLS Friends,

I began in April in the Neuromuscular Disorders Program as the "PLS Nurse". I am very pleased and excited to be a part of this program. I've come from a long and rewarding experience with the NIH-funded Women's Health Initiative (WHI), a group of studies for post-menopausal women. At our local site here at Northwestern, my responsibility was, along with my team, to maintain contact and continue collecting data from 1700 participants throughout the years of their involvement with the research. I expect that my experience with the WHI study will be a helpful background in expanding the Primary Lateral Sclerosis Registry while I continue at the same time to learn more about the intricacies of PLS. I am realizing how very difficult it is to arrive at the PLS diagnosis, how much we still have to learn.

I'm grateful to have had contacts with a number of PLS friends who have sent recent email queries to the websites and I am sending blood sample kits (around 4-6 per month, which I hope will soon double) to all who are interested in participating in the Primary Lateral Sclerosis Registry. To date we have blood samples from 184 who were thought to have a diagnosis of PLS at the time of our initial contact. A few of these have changed, and are now thought to be HSP (spastic paraplegia) or other neuromuscular disorders.

I am also contacting registry members to update contact information and medical records and to request completion of missing bits of information. Refining the collection and shipping procedures for spinal fluid (CSF) samples is another current project. Five CSF samples for PLS proteomic studies have now been received and 3 more are expected this summer. We have also decided to accept CSF samples even if a person has a Baclofen pump. Further plans for outreach to increase participation in the registry will include contacts with the neurologists who participated in the International PLS Meeting, then other neurologists in university settings in the US and Canada, then overseas.

I'd also like to mention that neurologists who have authored papers on PLS have emphasized the research value of examination of tissue after death. Even though that time is likely well into the future, several of you have already signed an "Intent for Autopsy Form" and have made arrangements in your living will for tissue donation to the Neuromuscular Program here at Northwestern. If others are interested in participating, I can provide more information.

Grace Carlson-Lund, RN
Clinical Research Nurse

Neuromuscular Disorders Program
Northwestern University Feinberg School of Medicine
303 East Chicago Avenue
13-715 Tarry Building
Chicago, IL 60611
Phone: 312-503-0160
Fax: 312-908-0865
Email: gcarlsonlund@northwestern.edu
Website:
www.neurogenetics.northwestern.edu

World Community Grid

Some in our patient community have joined the WCG. World Community Grid's mission is to create the world's largest public computing grid to tackle projects that benefit humanity. Our work has developed the technical infrastructure that serves as the grid's foundation for scientific research. Our success depends upon individuals collectively contributing their unused computer time to change the world for the better.

World Community Grid is making technology available only to public and not-for-profit organizations to use in humanitarian research that might otherwise not be completed due to the high cost of the computer infrastructure required in the absence of a public grid. As part of our commitment to advancing human welfare, all results will be in the public domain and made public. Grid technology is simple and safe to use. To start, you download and install a small program or "agent" onto your computer. When idle, your computer will request data on a specific project from World Community Grid's server. It will then perform computations on this data, send the results back to the server, and ask the server for a new piece of work. Each computation that your computer performs provides scientists with critical information that accelerates the pace of research!

Please go to http://www.worldcommunitygrid.org/viewJoinNow.do to learn more about the WCG and download the software if you would like to participate.

Protein Analysis of ALS Mice Spinal Fluid May Help Study of PLS Through PLS Registry

Contributed by Mark Weber

The SPF funded the PLS Registry in Dr. Teepu Siddique's lab at Northwestern University School of Medicine. The major goal of the project was to collect enough spinal fluid and blood from PLSers to be able to run experiments on those fluids to look for abnormalities specific to PLS as a way to begin to understand what is causing adult onset, sporadic PLS. What can be studied with mice spinal fluid can be studied with that of humans. And it can also be studied with Primary Lateral Sclerosis specimens. But we need more PLSers to donate before this crucial work can begin. Please see the letter from Grace L. Carlson-Lund, the nurse in charge of the registry.

Here’s a brief summary of the work with ALS mice.

SOURCE: Mol Cell Proteomics. 2006 Mar 29; [Epub ahead of print]
http://www.mcponline.org/cgi/reprint/M500431-MCP200v1

We did a protein analysis on spinal fluid on a line of mice. These mice have a mutated, human gene known to cause familial ALS in humans (and ALS symptoms in the mice). We compared it to spinal fluid from normal mice. We found protein abnormalities in the ALS mice. Some abnormalities occurred before symptoms appeared-others later on.

This information may lead researchers to discover what is causing the abnormalities and to be able to one day track the usefulness of treatments.

Lukas TJ, Luo WW, Mao H, Cole N, Siddique T. Molecular Pharmacology & Biol. Chemistry, Northwestern University, Chicago, IL 60611.

Research Snippets

Neurons Grown from Embryonic Stem Cells Restore Function in Paralyzed Rats

Source: *Annals of Neurology* July 2006

For the first time, researchers have enticed transplants of embryonic stem cell-derived motor neurons in the spinal cord to connect with muscles and partially restore function in paralyzed animals. The study suggests that similar techniques may be useful for treating such disorders as spinal cord injury, transverse myelitis, amyotrophic lateral sclerosis (ALS), and spinal muscular atrophy. The study was funded in part by the NIH's National Institute of Neurological Disorders and Stroke (NINDS).

The researchers, led by Douglas Kerr, M.D., Ph.D., of The Johns Hopkins University School of Medicine, used a combination of transplanted motor neurons, chemicals capable of overcoming signals that inhibit axon growth, and a nerve growth factor to attract axons to muscles.

31st HSP Gene Locus Discovered

The 31st HSP gene locus was recently discovered by a team of scientists from Duke University. The new gene locus is for a dominant gene and is located on chromosome 2. An article announcing the discovery was published in the journal Neurogenetics. The scientific team included Drs. Steven Zuchner, Melanie Kail, Martha Nance (the chairman of the SPF's Scientific Advisory Board), Perry Gaskell, Ingrid Svenson, Douglas Marchuk (recipient of an SPF research grant in 2003), Margaret Pericak-Vance, and Allison Ashley-Koch.

Single Mutation Identified that Leads to Motor Neuron Death

SOURCE: J Cell Biol. 2006 Feb 27;172(5):733-45.

http://tinyurl.com/lhhj2

A single point mutation in a particular gene causes both a loss of normal protein
function, and a resulting toxic protein. Together they lead to motor neuron death.

Levy JR, Sumner CJ, Caviston JP, Tokito MK, Ranganathan S, Ligon LA, Wallace KE, LaMonte BH, Harmison GG, Puls I, Fischbeck KH, Holzbaur EL. Department of Physiology, School of Medicine,
University of Pennsylvania,
Philadelphia, PA 19104, USA.

Botox Controls Sialorrhea (Excessive Drooling) in ALS

SOURCE: Muscle Nerve. 2006 Mar 31; [Epub ahead of print]

http://tinyurl.com/g46c6

Verma A, Steele J. Department of Neurology, Kessenich Family MDA-ALS Center, 1150 NW 14th Street, Suite 701, University of Miami Miller School of Medicine, Miami, Florida 33136.

Sialorrhea (excessive drooling) is frequently a socially disabling symptom in patients with bulbar amyotrophic lateral sclerosis (ALS). In this open-label prospective study, we report the effect of botulinum toxin A (Botox) injection into the parotid glands in 10 patients with bulbar ALS and socially disabling sialorrhea. We applied three different outcome measures to determine the effect of Botox therapy on sialorrhea. Botox significantly improved the degree of sialorrhea and a drooling impact score and, by inference, the quality of living, in over half of the patients with bulbar ALS and severe sialorrhea. The beneficial effect of Botox lasted for at least 2 months in those who responded. No major adverse effects were noted. Local injection of a small dose of Botox into the parotid glands can control sialorrhea and potentially improve living quality in some patients with bulbar ALS.

Adult Stem Cell Transplants Improve Recovery In Animal Models For

Stroke, Cerebral Palsy

http://www.sciencedaily.com/releases/2006/04/060408234948.htm

A single dose of adult donor stem cells given to animals that have neurological damage similar to that experienced by adults with a stroke or newborns with cerebral palsy can significantly enhance recovery. Using a commonly utilized animal model for stroke, researchers administered a dose of 200,000-400,000 human stem cells into the brain of animals that had experienced significant loss of mobility and other functions. The stem cells used in the study were a recently discovered stem cell type, referred to as multipotent adult progenitor cells, or MAPCs. Treated animals experienced at least 25 percent greater improvement in motor and neurological performance than controls, said Dr. Cesario V. Borlongan, neuroscientist at the Medical College of Georgia and the Veterans Affairs Medical Center in Augusta. The findings were presented April 7 during the 58th annual meeting of the American Academy of Neurology in San Diego.

Animal Model of HSP Created in Fruit Flies

SOURCE: http://www.fsp-info.de/sympos_06/abstract_sigrist1.pdf

Carola B. Sigrist, Frank F. Madeo* and Stephan J. Sigrist European Neuroscience Institute, Grisebachstr. 5, 37077 Göttingen *IMB, Universitätsplatz 2, Karl-Franzens Universität Graz, Austria

Dr. Sigrist and his colleagues have created a model of HSP in fruit flies by eliminating their Atlastin gene (An Atlastin knockout.). (The Atlastin gene, when mutated, causes juvenile onset HSP in humans.) In the absence of Atlastin, most fruit flies eggs could not produce live fruit flies. Those that did produced small, sterile fruit flies that showed severe movement problems. They also have significantly shorter lifespans than normal fruit flies.Dr. Sigrist and his team will now look for Atlastin binding partners, as well as the areas inside neurons where lots of the Atlastin protein is found.

Respiratory Function & PLS

SOURCE: Amyotroph Lateral Scler Other Motor Neuron Disord. 2006 Mar;7(1):57-60.
http://tinyurl.com/q88u2

Gouveia RG, Pinto A, Evangelista T, Atalaia A, Conceicao I, de Carvalho M.
Department of Neurology, Neuromuscular Unit, Hospital de Santa Maria, Lisbon, Portugal.

Primary lateral sclerosis (PLS) is a very rare disease characterized by pure upper motor neuron findings. Although a number of previous reports have evaluated this condition, no study has addressed the respiratory function in PLS. Six patients meeting previously proposed diagnostic criteria for PLS were submitted to a number of respiratory tests: forced vital capacity, maximal pressures, phrenic nerve responses, needle electromyography of the respiratory muscles, percutaneous nocturnal oximetry (PNO) and polysomnography (two patients). Our results show that the diaphragm is not affected in this condition, but some respiratory function tests (RFT) and PNO had abnormal values. Voluntary muscular activation to perform RFT may be limited in these patients. PNO and polysomnography suggest that respiratory central drive dysfunction can occur when upper motor neurons are severely affected, in PLS. However, we did not verify progression on follow-up.

Drug to Control Emotional Lability

SOURCE: J Neurol Sci. 2006 May 2; [Epub ahead of print] http://tinyurl.com/pyqnf

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Technion, Haifa, Israel.

Pseudobulbar affect (PBA), a condition involvinginvoluntary and uncontrollable episodes of crying and/or laughing, occurs frequently in patients with a variety of neurological disorders, including amyotrophic lateral sclerosis (ALS). Although PBA results in considerable distress for patients and caretakers, it is underrecognized and undertreated.

Dextromethorphan, a common cough suppressant, specifically targets sigma(1) receptors concentrated in the brainstem and cerebellum, thus providing the possibility of targeting regions implicated in emotional expression. When administered in a fixed combination with quinidine, dextromethorphan is effective in treating PBA in patients with ALS.

Management of Childhood Spasticity: a Neurosurgical Perspective.

SOURCE: Pediatr Ann. 2006 May;35(5):354-62.

http://tinyurl.com/rqk76

Mandigo CE, Anderson RC.

Department of Neurological Surgery, The Children's Hospital of New York, Columbia University, New York, NY 10032, USA.

The management of childhood spasticity requires a multidisciplinary effort. With input from pediatricians, physical and occupational therapists, neurologists, orthotists, orthopedic surgeons, neurological surgeons, and other healthcare personnel, effective treatment for spasticity can be initiated and maintained that can lead to meaningful improvements in quality of life for vast numbers of children. Neurosurgical treatment of spasticity will continue to evolve and be refined as procedures and techniques are appropriately evaluated with reliable and validated outcome measures.

The Role of Mitochondria in Inherited Neurodegenerative Diseases.

SOURCE: J Neurochem. 2006 Jun;97(6):1659-75.

http://tinyurl.com/p74en

Kwong JQ, Beal MF, Manfredi G.

Department of Neurology and Neuroscience, Weill Medical College of Cornell University, New York, New York, USA.

In the past decade, the genetic causes underlying familial forms of many neurodegenerative disorders, have been elucidated. However, the common pathogenic mechanisms of neuronal death are still largely unknown. Recently, mitochondrial dysfunction has emerged as a potential 'lowest common denominator' linking these disorders. Understanding the contribution of mitochondrial dysfunction to neurodegeneration and its pathophysiological basis will significantly impact our ability to develop more effective therapies for neurodegenerative diseases.

Monozygotic Twins Discordant for Primary Lateral Sclerosis

SOURCE: Amyotroph Lateral Scler Other Motor Neuron Disord. 2006 Jun;7(2):123-125.

http://tinyurl.com/lvyct

Sorenson EJ.

Department of Neurology, Mayo Clinic, Rochester, MN,US.

The genetics of the motor neuron diseases are becoming increasingly important. The relationship of primary lateral sclerosis and amyotrophic lateral sclerosis remains debated. Here a pair of monozygotic twins discordant for primary lateral sclerosis is presented. This monozygotic twin pair suggests that environmental factors were most relevant to this subject's development of primary lateral sclerosis. Their life history suggests very similar social and professional exposures without any apparent distinctions.

The Role of Alsin in PLS/HSP

Alsin/Rac1 signaling controls survival and growth of spinal motoneurons.
SOURCE: Ann Neurol. 2006 Jun 26;60(1):105-117 [Epub ahead of print] http://tinyurl.com/mefz4
Jacquier A, Buhler E, Schafer MK, Bohl D, Blanchard S, Beclin C, Haase G.
Institut National de la Sante et de la Recherche Medicale, Institut de Neurobiologie de la Mediterranee, Equipe AVENIR, France.
Recessive mutations in alsin cause juvenile amyotrophic lateral sclerosis (ALS2) and related motoneuron disorders. Alsin function in motoneurons remained unclear because alsin knock-out mice do not develop overt signs of motoneuron degeneration. Our data demonstrate that alsin controls the growth and survival of motoneurons in a Rac1-dependant manner. The strategy reported here illustrates how small interfering RNA electroporation can be used to generate cellular models of neurodegenerative disease involving a loss-of-function mechanism.

Progressive Spinal Axonal Degeneration and Slowness in ALS2-deficient Mice

SOURCE: Ann Neurol. 2006 Jun 26;60(1):95-104 [Epub ahead of print]
http://tinyurl.com/lnp6u
Yamanaka K, Miller TM, McAlonis-Downes M, Chun SJ, Cleveland DW.
Ludwig Institute for Cancer Research and Department of Medicine and Neurosciences, University of California, San Diego, La Jolla, CA.
Homozygous mutation in the ALS2 gene and the resulting loss of the guanine exchange factor activity of the ALS2 protein is causative for autosomal recessive early-onset motor neuron disease that is thought to predominantly affect upper motor neurons. The goal of this study was to elucidate how the motor system is affected by the deletion of ALS2. The combined evidence from mice and humans shows that deficiency in ALS2 causes an upper motor neuron disease that in humans closely resembles a severe form of hereditary spastic paralysis, and that is quite distinct from amyotrophic lateral sclerosis.

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PHOTOS

SP Connection with Reyerse Family in Washington, D.C.

Annette at dinner with Reyerse family

The Campbells with the Reyerses

Joanne Reyerse with Jim who is keeping up Frank's site

Living with HSP/PLS

NIH - Tracy, Thurza & Dr. Mary K. Floeter

Improvised handicapped transport at Dulles Airport

Flora Brand with “baby stroller”