Synapse

Summer 2003

Serving the Primary Lateral Sclerosis Community since 1997

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EVENTS & MEETINGS

Symposium Update

We did it! Our small group of PLSers has raised the $25,000 we needed before Dr. Siddique could apply to NIH for sponsorship of the Symposium. You'll clearly see the thermometer now at $31,456! The additional funds will go toward covering some other Symposium budget items. Dr. Siddique's office now is finalizing the dates and location for the Symposium. It will be held in Spring, 2004. We expect to be able to give you more specifics very soon. Thanks to all who donated personally and to those who worked hard to solicit donations from family, friends and business associates.

TeamWalk for the Cure
September 12-14 Lexington, MA

The **TeamWalk For the Cure** walkathon is the annual event of the SP (Spastic Paraplegia) Foundation to raise funds to find the cures for the upper motor neuron disorders Primary Lateral Sclerosis and Hereditary Spastic Paraplegia. The TeamWalk will be 10:00 a.m., September 14 in historic Lexington, Massachusetts. It will launch with a ceremony performed by members of the Minute Man Company and pass by Revolutionary War attractions including the Minuteman Headquarters. It all takes place during our glorious fall foliage season. Make travel plans early - flights and hotel rooms sell out!

If you can't attend the TeamWalk, then join the team from afar as a Walker by Proxy and a designated Walker will carry your name and spirit. We'll also post your photo on our Walker by Proxy Photo Board. Or organize a walk, swim or run at your home that day. Either way, you'll raise sponsorships for your participation in the TeamWalk, sending critical dollars to research.

The weekend also features our national "**Research and Beyond" Conference**, featuring keynote speaker Robert Brown, M.D., Mass General Hospital, a leading expert on neurodegenerative disorders. His program will feature cutting edge research and treatment for PLS and HSP. There's also a "Walking Straight" program as well as special sessions and social events both Friday and Saturday evenings.

Register online by August 22 www.sp-foundation.org or if you receive the print version of Synapse, register using the enclosed form.

Last year's TeamWalk raised $80,000 for research! One $40,000 grant is being awarded for PLS and one for HSP. Let's aim for $100,000 this year.

Our Great Day Today at the FL Connection

Contributed by Jane McCord

What a fun day we had June 7! Thanks to Carol Liquori and Ann White, 19 of us got acquainted or reacquainted with other PLS and HSP men and women and their caregivers. Joe Alberstadt (who, for the newcomers, started the PLS-FRIENDS and Synapse several years ago) and his wife, Carol, attended and it was great seeing them again. We had several speakers: a demonstration of several power chairs or scooters, which all those interested drove around and had lots of fun doing this; an interesting talk by a representative from Lifeline Medical Alert; an excellent presentation on the Intrathecal Baclofen Pump; a very good talk about Theft Identity and how to avoid it and a talk about The Great Balancing Act - "Avoiding Falls." These were all done well. But in-between we had plenty of time to socialize, too.

The food, all made and catered by Sandi Burns, was delicious and included a wonderful breakfast, a lovely lunch and afternoon snacks.

All in all, I think everybody would agree with me that they enjoyed the whole day. We hope to do it again and hope that others will be able to join in the fun at the next Florida PLS/HSP Connection!

Connecticut Connection June 21

Contributed by Dolores Carron

We had a nice time on Saturday. Our meeting was at the Wethersfield Country Club again. We were a congenial group of 12, five of whom were newcomers. The day began with Continental breakfast and informal chatting. Our morning speaker was Scott Dyson, an experienced and knowledgeable rehabilitation equipment consultant. His presentation included useful information/hints on the modalities and programs available for maintaining our independence and daily functioning. That theme continued during lunch with a great deal of sharing and exchange of concerns, experiences and ideas among the group members.

In advance of the meeting, I had requested participants to submit written questions/concerns for discussion. I acted as moderator and introduced the submitted topics for group discussion. The sunny day ended with afternoon showers. All would agree that it was nice to see old friends and to make some new ones. We look forward to the next time.

Phoenix Connections June 21

Compiled from reports by Bonnie Guzelf and Kathi Geisler

The hot air balloon ride was fabulous. The weather was delightful and the vistas interesting and the sunrise was lovely. A great ride! We enjoyed mimosas and muffins and fruit then the van took us back to the hotel. We got back to the hotel about 8:00 a.m. (!!!!) Great way to start the day.

A delightful Hospitality Room was set up for Friday arrivals. 30 people attended from all over the country and Canada. Reese the Wonder Dog was the big hit. Reese is Michael Breaux's helper dog. Michael has had juvenile PLS since he was 13 and Reese goes everywhere with Michael.

The Connections was fabulous - a great combo of learning, sharing and fun. On Saturday we had a lovely luncheon at the hotel. Fay Fishman and Dennis Peterson Attorney's at Law spoke for about an hour on the subject of Disability Insurance, both Social Security and Private. Then Kiera Widmer lead us in some chair Yoga stretches and breathing. Bonnie did a short guided meditation for relaxation. Then several students from the local massage school came in and offered FREE back and shoulder massage. What a nice way to end the day.

Several of us enjoyed a great Mexican restaurant after the Connections for great food and conversation. "I (Bonnie) wish it could have lasted longer. It was so nice to meet new friends and to put faces on some of the names. It's so empowering to meet others who deal with the same challenges every day."

Ann Arbor Support Meetings

Joan Mathay, Dr. Fink's nurse, runs a monthly support group meeting held at the University of Michigan Cancer Center. The attendance is almost all PLS/HSP, although it is actually started out as a "NORD" group.

The size varies from a few people in attendance to a good handful. Every meeting has a topic of discussion and sharing. It is held every second Friday of the month, 6:30 pm - 8:30 pm. For more information, please contact Joan Mathay:
734-936-3087
jmathay@umich.eduA

October 17-19 Forum in Washington, DC

Contributed by Kathi Geisler

WASHINGTON, DC - The American Association of People with Disabilities (AAPD) and the National Organization for Women (NOW) Foundation announce that they will hold a joint forum on disability and women's rights October 17-19, 2003. Entitled Women with Disabilities & Allies Forum: Linking Arms for Equality & Justice for All, the three-day summit will address issues of mutual concern to the feminist and disability rights movements.Go here for more information: http://www.aapd-dc.org/docs/NOWwomanforum.html

If you are interested in participating in the forum or a connections meeting in DC, please email me so we can coordinate something : kathi@sp-foundation.org or kathigeisler1@aol.com.

FUNDING

Swimming Fundraiser for PLS was Great Success!

The Bulldog Aquatic Club (BAC) is a United States Swimming team of youth up to age 18. Ours is a relatively small team - approximately 100 families. For most all of those 22 years, the club generated operating funds solely by hosting one (USS) sanctioned swim meet annually. Because our club has provided such a quality meet in recent years, the governing Michigan swimming organization asked our club to consider hosting the Michigan State Championship meet for age 13 and over, this spring. These discussions/considerations began last summer - the time at which BAC board members began to learn of Kurt Ohlgren's diagnosis of PLS.

And so we ran the state meet. We directed the "Bulldog Challenge Fundraiser" to the top 7 USS officials in charge of the meet. I chose not to make the fundraiser an open effort to all clubs but rather a confidential effort aimed specifically at the people outside our club directly knowledgeable/aware of Kurt's diagnosis. There was no mention of this "Challenge" in the program but rather I mailed individual invitations to the officials, to join BAC in contributing to the PLS Symposium Fund.
The meet itself was held at Eastern Michigan University in Ypsilanti, MI from March14-16, 2003. 565 athletes attended from a total of 49 clubs across Michigan. We were able to donate $1075.00 to the PLS fund.

Raffle Time Again

PLSer Donna Isenhour has made beautiful splarkling one-of-a-kind poinsetta cafe curtains with a valance - and you have a chance to win them. To see the curtains, go to the Donor Opportunities section of the Synapse site. Chances are 6 for $10.00. Send a Check made out to **NUMS/PLS Symposium** in increments of $10.00 along with your name and address to:
Donna Isenhour
2882 Palmer Dr.
Conover, NC 28613
Donna will put the correct number of chances into the "pot" for you. The drawing will take place in the fall - in plenty of time for Christmas. The curtains will be on display at TeamWalk Sept. 12-14.
Donna has also finished three more curtain toppers: apple blossom, sunflower and grape clusters. You may order the curtain topper of your choice from Donna for $38. each. All of the money from these curtain toppers will also go to NUMS/PLS Symposium.

MEDICAL UPDATES

Primary Lateral Sclerosis

Authors: Doctor Frans Brugman and Professor John Wokke. Scientific Editor: Professor Marianne de Visser Orphanet Encyclopedia. December 2002:
http://orphanet.infobiogen.fr/data/patho/GB/uk-PLS.html
Ed. Note: the article below has been adapted by me for Synapse with approval of Dr. Brugman
Diagnostic criteria / definition PLS is an idiopathic non-familial neurodegenerative disorder of upper motor neurons. PLS presents clinically as a slowly progressive pyramidal tract syndrome, sometimes with marked pseudobulbar symptoms. The diagnosis is based on exclusion of other causes and requires normal findings of blood, urine and CSF tests, normal EMG findings and no abnormalities of brain and spinal cord MRI. In 1992, Pringle and colleagues proposed diagnostic criteria for PLS. These criteria are generally accepted and consist of adult onset of symptoms and a negative family history, in addition to normal findings of the above mentioned additional tests. The Pringle criteria require a disease duration of at least three years to exclude ALS, as the initial presentation of ALS can be a pure upper motor neuron syndrome. . However, transition to ALS has been described in patients with longstanding PLS, even after a disease duration of as long as 18 years.
- Primary progressive multiple sclerosis (PPMS) can show a pure motor syndrome.
- ALS can initially present as a pure upper motor neuron syndrome, before the appearance of clinical or neurophysiological signs of lower motor neuron loss.
Frequency PLS is very rare. The incidence of PLS can be estimated at approximately a little less than 1 per 1,000,000 a year. The prevalence is probably around 10-20 per 1,000,000, since PLS has a chronic disease course.
Clinical description PLS is characterized by a slowly progressive bilateral pure upper motor neuron syndrome. Disease onset is usually between 40 and 60 years with spasticity in the legs. Onset in the pseudobulbar region with speech and swallowing disturbance is also possible. Ultimately a tetrapyramidal syndrome develops, sometimes with marked pseudobulbar

features (forced laughing and crying). Disease progression is usually very slow over many years. Life expectancy is normal. Considerable functional impairment can develop over time. Bladder involvement, in the form of urgency and frequency symptoms, occurs in approximately half of the patients, usually late in the disease course. Subclinical neuropsychological disturbances have been described in PLS patients in some studies, but dementia is exceptional.

Management including treatment There is no cure for PLS. Treatment of PLS is symptomatic and can involve prescription of antispasticity medication and rehabilitation.

Etiology The etiology of PLS is not known. Since the original description by Erb in 1875, it has long been debated whether PLS exists as a disease entity. PLS is now considered to be a benign variant of ALS, at the one end of the clinical spectrum where there are only upper motor neuron symptoms

Unresolved questions

Many questions are currently unresolved:

- Does PLS represent a single disease entity or a group of entities?
- Are there any prognostic factors and/or (genetic or other) susceptibility factors?
- What proportion of patients develop signs of anterior horn cell disease?
- What proportion of patients show progression to ALS, and how does this disease evolution affect the prognosis?
- Does riluzole, a drug with a small life-prolonging effect on the survival of ALS patients, have also an effect on the disease course in PLS?

A Comparison of Upper and Lower Motor Neuron Physiology in Primary Lateral Sclerosis (PLS) and Amyotrophic Lateral Sclerosis (ALS)

Stewart H, Eisen A, Weber M Neuromuscular Diseases Unit, Vancouver General Hospital, Vancouver, Canada; Department of Neurology, Kantonsspital St Gallen, St. Gallen, Switzerland

Background and objectives

The majority consensus is that PLS forms one end of a clinical spectrum of ALS, characterized by slow progression and restriction to upper motor neuron deficits for long periods. PLS is rare, and comparative physiology with ALS is sparse. Clues to the mechanism underlying slow progression in PLS might be of benefit in ALS.

Methods

We have compared corticomotor neuronal (upper motor neuron) and anterior horn cell (lower motor neuron) function in PLS *(n* 10), ALS *(n* 15), and age-matched normal controls *(n* 15). Control subjects were neurologically normal, and PLS patients had no clinical evidence of lower motor neuron disease at the time of study. Motor unit number estimates (MUNES) from the thenar muscle group were used to investigate lower motor neuron function.

Standard electromyography (EMG) of the abductor pollicis brevis muscle was also performed. Motorevoked potentials (MEPs), central motor conduction time (CCT), and peristimulus time histograms (PSTHs) resulting from transcranial magnetic stimulation (TMS) were used to investigate upper motor neuron function. PSTHs were con-structed from 4—5 different, voluntarily recruited motor units in each patient and the onset latency, number of excess bins, duration, amplitude and synchrony of the primary peak (PP) were measured.

Results

Thenar MUNES were significantly reduced in ALS (mean 67 55) compared to normal controls (mean 256 76, *P* 0.001). Thenar MUNES were also reduced in PLS (mean 125 55, *P* 0.05) compared to normal controls, although thenar muscle wasting was not present, MRC strength was normal, and standard EMG of the thenar muscle group was unremarkable in these patients. In 50% of PLS patients, a MEP could not be evoked. The cortical threshold in the remaining 5 PLS patients was 79.4 14.4%, significantly higher *(P* 0.01) than in ALS (60.3 12.9%) and normal controls (51.4 9.5%). Central motor conduction was 11.6 3.1 ms in PLS, 7.2 2.4 ms in ALS, and 6.7 2.6 ms in normal controls. Single motor unit responses were abnormal in PLS and ALS as

compared with normal controls, with higher stimulus thresholds, and delayed, desynchronized PPs with prolonged durations. These abnormalities were most striking in the PLS group.

Conclusion

The major physiological differences between PLS and ALS are a relative absence of EMG abnormalities, high cortical threshold to TMS, and slowed central conduction to both a whole target muscle and a single motor unit. However, MUNES were reduced in PLS as well as ALS, despite the lack of lower motor neuron disease in PLS. Different mechanisms are likely to be involved. In ALS, anterior horn cells are truly lost, whereas in PLS, the reduced MUNE might reflect decreased excitability of some motor axons. This may be a consequence of chronic, inadequate cortical drive to the corresponding anterior horn cell, rendering it dysfunctional as opposed to dead. Transsynaptic cell degeneration may explain the conversion of many patients with PLS to ALS. The differences found in this study are not absolute, and variable overlap probably occurs in both syndromes. Functional loss of motor units implies the potential for reversibility.

Primary Lateral Sclerosis: A heterogeneous disorder composed of different subtypes?

P. Zhai, MD, PhD; F Pagan, MD; J Statland; JA Butman, MD, PhD; MK Floeter, MD,PhD. NEUROLOGY, April, 2003; 60:1258-1265.

Twenty-five patients meeting previously proposed diagnostic criteria for PLS were seen for examination, measurement of gait and finger tapping speed, and various tests to assess motor neuron pathways. The excitability of the motor cortex, and central motor conduction time were assessed with transcranial magnetic stimulation (TMS). Brainstem motor pathways were assessed by the acoustic startle reflex. Magnetic resonance spectroscopy (MRS) was performed on a group of patients to assess metabolites in the motor cortex.

Results: Startle reflexes were often hyperactive in PLS patients, but mostly in patients with speech difficulties of recent onset.

56% of patients had a similar pattern of symptom progression—termed ascending PLS. In those patients, spasticity began in the legs, and progressed slowly and steadily. Spasticity in the arms developed 3.6 years after the legs, on average (range 1-6 years).

Speech impairment followed an average of 1.5 years later (range 0.5 – 10 years).

This pattern of progression is compatible with a steady dying back of the corticospinal axons. A similar length dependent axonopathy has been proposed for some forms of HSP. (Fink, 2001.)

In the group of ascending PLS patients, all patients had elevated TMS threshold with absent motor evoked potentials—a finding that can indicate either loss of cortical neurons, or terminal axonal degeneration. Loss of motor evoked potentials is similar to findings late in ALS.

To determine whether PLS patients in the ascending group had a loss of cortical neurons, MRI scans of the motor cortex were examined for areas of atrophy. No areas of atrophy were found compared to age matched (normal) controls. But MRS showed reduced NAA/Cr ratios in the arm area of the motor cortex of these patients compared with controls, indicating neuronal atrophy, dysfunction, or loss. Similar reductions in NAA/Cr ratios have been described in ALS. Experimental models have shown that declining NAA/Cr ratios may reflect neuronal inactivity rather than loss of neurons. Thus it remains uncertain whether motor cortex neurons have been irreversibly lost in patients with ascending PLS.

Patients not in the ascending group showed differing types of symptom progression, with little correlation between symptom progression and test results.

The goal of this study was to identify subsets of patients with PLS with common clinical, physioloical, and anatomical features, as a first step in investigating causes of PLS. Patients with ascending PLS may offer a better sample for

genetic investigations compared to the overall PLS population—as PLS is currently defined.

Clinical trials at NIH

Contributed by Jennifer Thompson

This site, http://clinicaltrials.gov/search/term=Primary+Lateral+Sclerosis lists all trials which relate to PLS. Go to the site for details about participating.

1. Recruiting Nuclear Magnetic Spectroscopy Imaging to Evaluate Primary Lateral Sclerosis, Hereditary Spastic Paraplegia and Amyotrophic Lateral Sclerosis

   Conditions: Primary Lateral Sclerosis; Hereditary Spastic Paraplegia; Amyotrophic Lateral Sclerosis

2. Recruiting Physiologic Studies of Spasticity

   Conditions: Muscle Spasticity; Healthy

3. Not yet recruiting A Multi-Center Phase III Trial of Minocycline in Amyotrophic Lateral Sclerosis

   Condition: Amyotrophic Lateral Sclerosis

4. Not yet recruiting IGF-1/ALS Trial

   Condition: Amyotrophic Lateral Sclerosis

5. Recruiting Clinical Trial of Creatine in Amyotrophic Lateral Sclerosis

   Condition: Amyotrophic Lateral Sclerosis

6. Recruiting Safety testing of AVP-923 in the Treatment of Emotional Lability (Uncontrolled Laughter & Crying)

If you do not have computer access, send your inquiry to:

National Institutes of Health
Dr. Mary Kay Floeter
9000 Rockville Pike
Bethesda, MD 20892

ALS Therapy Development Foundation

http://www.als.net/alstdf/treatments/ is a site which should be a favorite. You will find the latest research information here, as well as detailed information on drug treatments – both prescription and over the counter. Many neurologists feel that what helps classic ALS probably helps PLS, too.

New Technology May Help us All

Contributed by Mark Weber and Katherine Wisehart

There is a new, experimental technology that may have great promise from PLSers and HSPers. The technology is called "tensor diffusion imaging" or "tensor diffusion MRI. Although still very much experimental, there is hope that this technology may be able to detect damage to nerve cells in a way that other imaging technologies (x-ray, CT scan, conventional MRI) cannot. Scientists are already using the technology to detect damage to neuronal axons in MS patients. For more info about this technology, go to PubMed at http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?db=PubMed and search for "tensor diffusion spinal cord" and "tensor diffusion cortex". (Put the words in quotes so you don't get lots of useless articles.)

Update from NINDS @ NIH

Contributed by Stephanie Clipper Public Liaison Officer NIH Neurology Institute (NINDS-NIH-DHHS) NINDS notes are now available on-line: http://www.ninds.nih.gov/funding/nindsnotes_06_03.htm and contain information about clinical trials in recruitment, new initiatives and other items of interest to PLSers

"The Word on Health," which is a free consumer health publication of the National Institutes of Health is also available: http://www.nih.gov/news/WordonHealth/

For the latest in lay language on genes, go to http://ghr.nlm.nih.gov .

Stem Cell Strides Test Bush Policy

By Rick Weiss, Washington Post

Contributed by Jeff Glassman; condensed by Editor

A series of important advances have boosted the potential of human embryonic stem cells to treat heart disease, spinal cord injuries and other ailments, but researchers say they are unable to take advantage of the new techniques under a

two-year-old administration policy that requires federally supported scientists to use older colonies of stem cells.

Now pressure is building from scientists, patient advocates and members of Congress to loosen the embryo-protecting restrictions imposed by President Bush, with some on Capitol Hill saying they want to take up the issue.

Stem cells obtained from 5-day-old human embryos can morph into all kinds of human tissues and appear capable of regenerating ailing organs. But while newer and safer versions of the cells have recently been created, the policy imposed by Bush in August 2001 puts those cells off-limits to any scientist whose work is supported with federal money.

The older cells allowed under the Bush plan are less attractive to researchers because they have been grown in mixtures with mouse tumor cells. The tumor cells, known as "feeder" cells, secrete crucial, though as yet unidentified, nutrients that help nourish human embryo cells. But they can also pass mouse viruses or other microbes to the human stem cells, which means the stem cells could end up sickening patients instead of curing them. Concerned about that risk, the Food and Drug Administration has said it will demand an array of safety tests and long-term patient follow-up for any experiments in which patients are given stem cells that have been in contact with animal feeder cells.

Recently, however, scientists have learned how to grow human embryonic stem cells without mouse cells. The new stem cell colonies, or "cell lines," appear ideal for use in clinical trials, scientists say. But they remain unavailable to the vast majority of U.S. stem cell researchers -- most of whom depend on federal grant money -- because the 2001 Bush policy requires those scientists to work only with cells from embryos destroyed before Aug. 9, 2001. The goal was to prevent further destruction of stored human embryos (in fertility clinics), but it also limits researchers to cell lines tainted by contact with mouse cells.

"This is the conundrum we're caught up in as federally funded researchers under the Bush policy," said George Daley, a Harvard University stem cell biologist also affiliated with the Whitehead Institute for Biomedical Research in Cambridge, Mass. "We want to do the basic research that works towards cures, but we cannot use the newly derived, latest and best cell lines, which puts us at a disadvantage."

Foes of embryo cell research said they remain opposed to any changes. The White House indicated it has no intention of changing its position.

Clinical scientists and patients have been clamoring for a simpler approach. Scientists at Johns Hopkins University made a leap, showing that human embryonic stem cells could thrive in a culture system containing adult human bone marrow cells, which apparently secrete all the growth factors the stem cells need. The marrow cells offer "a clinically and ethically feasible method to vastly expand human embryonic stem cells on a clinical scale," concluded Linzhao Cheng and his colleagues in the March issue of the journal Stem Cells. "It's probable that many different human cell types can support the growth of [embryonic stem] cells in the right conditions," Cheng said. "It's certainly broader than we thought."

Thomas Okarma, president and chief executive of Geron Corp. of Menlo Park, Calif., said his company is close to having an all-human culture system for stem cells. With recent animal studies looking quite positive, he said, he could imagine human clinical tests beginning as soon as a year from now.

Lyme Disease 101 Part 2

Compiled by Sallie Longeri, Lyme Disease survivor

Lyme disease was named in 1977 when arthritis was observed in a cluster of children in and around Lyme, Connecticut.

Ticks are bloodsucking external parasites that feed on humans, wild and domestic mammals, birds, reptiles and others. They are totally dependent on the blood/tissue fluids of the host.

The longer an infective tick feeds, the greater the chance of infection.
Ticks are not insects. Ticks are arachnids, as are chiggers, spiders and mites. Ticks go through four life stages: egg, larva, nymph, and adult. The egg hatches into a larva. The larva feeds and molts into a nymph. The nymph then feeds and molts into an adult. The female requires a blood meal in order to lay eggs.Larvae and nymphs feed primarily on small mammals (especially the white-footed mouse, other rodents, and insectivores), and also on birds, dogs, deer, and humans. Nymphs aggressively bite humans. Adults feed primarily on deer, but also attach to large mammals (foxes, raccoons, opossums, dogs) and humans.
The adults attach to the white-tailed deer, engorge, and mate. The male dies after mating but the female continues to feed until egg development is completed and remains on the deer until spring when she drops off to lay eggs. Shortly after her eggs are laid, the female dies. Females lay up to 3000 eggs in soil and litter. The life cycle may range from 2-4 years and is regulated by host abundance and physiological mechanisms. Larvae are active from July through September, nymphs from May through August, and adults in the fall, winter, and early spring (October-May).
Ticks do not fly. They remain at ground level or slightly above by climbing onto vegetation. The tick achieves its blood meal by using its forelegs to feel/grab for a host. Ticks are usually found from ground level to three feet above the ground. A tick uses carbon dioxide, scent, body heat, and other stimuli to find a host. Nymphal deer ticks, the most common transmitters of Lyme disease, are often about 2mm.

Recent article on Stem Cell Research at Harvard

The full article may be found at the website below:
http://www.news.harvard.edu/gazette/2002/11.21/01-stem.html

LIVING WITH PLS

Friends

Compiled by Linda Gentner

True friendship is like sound health;
the value of it is seldom known until it is lost.
-- Charles Caleb Colton

A real friend is one who walks in when the rest of the world walks out.

Don't walk in front of me, I may not follow.
Don't walk behind me, I may not lead.
Walk beside me and be my friend.
-- Albert Camus

Strangers are just friends waiting to happen.

Friendship is one mind in two bodies.
-- Mencius

Friends are God's way of taking care of us.

I'll lean on you and you lean on me and we'll be okay
-- Dave Matthews

Everyone hears what you say.
Friends listen to what you say.
Best friends listen to what you don't say.

Hold a true friend with both your hands.;
-- Nigerian Proverb

A friend is someone who knows the song in your heart and can sing it back to you when you can't sing the words.

H.R.2700

Title: To amend title XVIII of the Social Security Act to revise the methodology by which payment for orphan drugs and biologicals is made under program prospective payment system for hospital outpatient department

services under the Medicare Program.
Sponsor: Rep Cox, Christopher [CA-48] (introduced 7/10/2003) **Cosponsors:** 9
Latest Major Action: 7/10/2003 Referred to House committee. Status: Referred to the Committee on Energy and Commerce, and in addition to the Committee on Ways and Means, for a period to be subsequently determined by the Speaker, in each case for consideration of such provisions as fall within the jurisdiction of the committee concerned. For up to date status of this bill which can affect PLSers, click on http://thomas.loc.gov/cgi-bin/query
Contact your US Representative to be sure they are in support of this bill.

Thoughts from Joe

Contributed by Joe Alberstadt – written in 1999 as he was leaving for Hawaii
Straight back loading on the plane doesn't hold me back. I hope my example will encourage you all to do all you can. Don't just sit back on your can. Have a great life . You can you know. Together we can all work to finding a link to a cause of PLS and ultimately a therapy. We can't do it by hiding in cyberspace. We have to trust each other to know who we are, where we live, how long affected, what do we suspect triggered etc. etc. Do it for yourself, do it for others, do it to help research and future generations. You think your little offering will not help "THAT'S BULL"

Long Term Disability

Contributed by Kathi Geisler, edited by Mark Weber
I learned something new at the Phoenix Connections that may be important to some of us regarding long term disability. For those of us who are working and paying into a long term disability insurance program, it is important to check into whether you are paying your policy premiums pre or post-tax.
The lawyer at the Phoenix Connections said that if you are paying your premiums pre-tax, that if you ever need to claim long-term disability, your benefits will be taxed. However, if you pay your premiums after-tax, then your long-term disability benefits will not be taxed. I am neither a CPA nor an accountant, so this is not professional advice from me or the SPF, but I am simply passing on something you may want to look into.
On the topic of disability, the lawyer also indicated that it is much easier to get Disability if you are over age 50. That is a magic number. That's because they don't figure you can be trained or educated for a new job after that age.

Medicare Patient Access to Drugs for Rare Diseases Act

Representative Chris Cox (R-CA), a member of the Energy and Commerce Committee and Chair of the House Select Committee on Homeland Security, introduced the Medicare Patient Access to Drugs for Rare Diseases Act of 2003 on July 10. The bill addresses concerns of the rare disease community as they relate to access to orphan therapies through Medicare. The full text of H.R. 2700 can be accessed at http://thomas.loc.gov. Then type HR2700 into the search box.
If you would like your representative to cosponsor as well, ask them to contact Sarah Petry, Legislative Assistant in Mr. Cox's office, to sign on. She can be reached at 202.225.5611.

Good Dieting Habits

Contributed by Steve Clark
I have been wheelchair bound 4 years this January. One of my biggest challenges was to not gain excessive weight. As of today, I weight a few pounds less than when I went in the chair. I too was very active and burned lots of calories with daily activities and exercise. My methodology to maintain and not gain is as follows:
1. Common sense when eating. You know what food is heavy in saturated fats and calories.
2. Go ahead and eat what you want, just eat a third or so of it.

3. Eat very slowly and take small bites and enjoy the food, eat and talk.
4. Do not eat and watch television, your brain and stomach have trouble communicating when it is focused on TV.
5. Eat 4 or 5 small meals during the day in lieu of 3 bigger ones as I was conditioned to do.
6. Obviously, drink some water, a little more than you probably do.
7. When I drink a diet caffeine free coke I add an equal amount of water. Sounds bad, but over time you get used to it and a straight coke will seem very strong.
8. Limit consumption amounts of any food that is WHITE, complex carbs, potatoes and pasta are my enemies. I know now why marathon runners consume massive quantities of pasta days before the race or as they say "carbing up." Without the race you will gain weight.
9. I have grown to love apples, bananas, plums, grapes, etc. Really good food and I can eat ALL I WANT!!
10. Green vegies are awsome, asparagus, fresh green beans, I like brussel sprouts and cabbage also.
11. A few don'ts from a wise doctor I know that is a cardiologist that he lives by:
A. Don't eat anything from a package that has been processed.
B. Don't eat anything from a "fast food restaurant."
C. Don't eat animal fat. You will get plenty of fat while trying to not eat fat. You need some fat obviously and you will get it as you will break down and eat some bacon or steak occasionally
12. Never eat in bed. Unless of course you are bedridden.
13. And last but not least, pay close attention to what goes in your mouth after 5 in the PM. Snacking is a killer. My snack is hot air popcorn.

My tips work for me, hopefully some will be helpful to you. It is a struggle but it can be done. EAT TO LIVE DON'T LIVE TO EAT!!

The Checklist to Live By

Contributed by Ronnie Grove
The most destructive habit.....Worry
The greatest Joy.....Giving
The greatest loss..... Loss of self-respect
The most satisfying work..... Helping others
The ugliest personality trait..... Selfishness
The most endangered species.....Dedicated leaders
Our greatest natural resource..... Our youth
The greatest "shot in the arm"...Encouragement
The greatest problem to overcome..... Fear
The most effective sleeping pill.....Peace of mind
The most crippling failure.....Excuses
The most powerful force in life..... Love
The most dangerous pariah..... A gossiper
The world's most incredible computer.....The brain
The worst thing to be without..... Hope
The deadliest weapon..... The tongue
The two most power-filled words..... "I Can"
The greatest asset..... Faith
The most worthless emotion..... Self-pity
The most beautiful attire.....SMILE!
The most prized possession.....Integrity
The most powerful channel of communication.....Prayer
The most contagious spirit..... enthusiasm
The most important thing in life..... GOD

Imagine...

There is a bank that credits your account each morning with $86,400. It carries over no balance from day to day. Every evening deletes whatever part of the balance you failed to use during the day. What would you do? Draw out ALL OF IT, of course!!
Each of us has such a bank. Its name is TIME. Every morning, it credits you with 86,400 seconds. Every night it writes off, as lost, whatever of this you have failed to invest to good purpose. It carries over no balance. It allows no overdraft.
Each day it opens a new account for you. Each

night it burns the remains of the day. If you fail to use the day's deposits, the loss is yours. There is no going back. There is no drawing against the "tomorrow". You must live in the present on today's deposits. Invest it so as to get from it the utmost in health, happiness and success! The clock is running. Make the most of today.
To realize the value of ONE YEAR, ask a student who failed a grade.
To realize the value of ONE MONTH, ask a mother who gave birth to a premature baby.
To realize the value of ONE WEEK, ask the editor of a weekly newspaper.
To realize the value of ONE HOUR, ask the lovers who are waiting to meet.
To realize the value of ONE MINUTE, ask a person who missed the train.
To realize the value of ONE SECOND, ask a person who just avoided an accident.
To realize the value of ONE MILLISECOND, ask the person who won a silver medal in the Olympics.
Treasure every moment that you have! And treasure it more if you shared it with someone special, special enough to spend your time.
Yesterday is history.
Tomorrow is a mystery.
Today is a gift.
That's why it's called the present!!

Cruising with PLS

Contributed by Bonnie Guzelf

We chose a Holland America cruise because my research showed that Holland America and Princess were the most "user friendly" for disabled people. We chose a new ship, the Zuiderdam, because the newer ships have been built to be accessible and have more accessible cabins available, while the older ones are do not. We got a handicap accessible cabin with a private balcony.The cabin was nice with a large bathroom with a shower for a roll in wheelchair. It had a fold down seat and a hand held shower.It was very convenient and I had no problems. There was even a ramp to get out to our balcony. The ship was totally accessible.

Although I have a jazzy power chair at home, I did not try to bring it with me. Instead, I rented a power chair from a company called Scoot Around America. They have local contacts in many cities. They delivered the chair to the ship (to my cabin) and picked it up when we left. I used it on board ship and it was wonderful! I did not have to depend on my husband to push me in a manual wheelchair. I would bring my chair to the dining room or showroom, then transfer to a regular seat and the staff would "park my buggy" until I needed it. If you can't transfer, no problem, the staff was very helpful and accommodating. I felt very independent. The price was $250 for the week, and well worth the money. We did bring my manual wheelchair along however, for Shore Excursions. This arrangement seemed to work very well. We took the chair with us to the airport and right up to the gate where we checked it (like a stroller). It was waiting for us when we got off the plane.

Shore Excursions: We did take several of the offered Shore Excursions. They worked ok, but in hindsight, I think it would have been better to just get a cab and go off on our own. The one trip that was memorable was the Dolphin Encounter in Nassau.

We were able to hug kiss and cuddle with the Dolphins. The people there were very helpful and I had no problems. I did learn one thing about Dolphins...When you rub a Dolphins tummy, do not rub him below the belly button.... or you will get a VERY EXCITED DOLPHIN. LOL!!!
What I learned, I think, was that I can still travel, I just have to put a little more thought and planning into it. I think that, for me at least, a cruise is a good way to travel. Everything is there for you and all (or most) of your needs are taken care of. It is important however, to chose the right ship and the right line.
Here are some travel websites to check out:
http://www.sath.org/
http://www.access-able.com/

http://www.mossresourcenet.org/travel.htm
http://www.emerginghorizons.com/
Thanks to David Lehman for these.

PLSers On Vacation

Bonnie Guzelf with Dolphin friend in Nassau

Connection attendees ready for pre-dawn launch in Phoenix

Sue Niquette parasailing with husband in Naussau

Synapse Editor Thurza Campbell (in six person paddle boat next to guide) on rafting trip with family in Idaho

Symptom & Medication Chart

Contributed by Jagan Baroda, PLSer in India

I find that keeping my own record of abilities is useful - 1) in focusing on my abilities from time to time. 2) In communicating with doctors / friends. 3) for tracking my progression. I have kept such a chart ever since I was diagnosed with PLS in 1991.

Enter a statement of fitness level from 1 to 10 for each symptom. **10** = totally **Fit**; **0** = absolutely **Unfit**

Symptom & Medication Chart

Parameter to Measure	**Year Diagnosed**	**Year +1**	**Year +2**	**Year +3**	**Year +4**
Age					
Weight					
Symptoms					
• Walking/balance					
• Speech					
• Breathing					
• Saliva					
• Swallowing					
• Exhaustion					
• Cramps/					
• constipation					
• Urinary urgency					
• Startling reflex					
Medications (name & dose)					
•					
•					
•					
•					
•					
Assists					
• Cane					
• Walker					
• Wheelchair					
•					

CAREGIVING

Caregivers Need Help

Summarized by editor from WebMD
http://my.webmd.com/content/Article/64/72500.htm

The daily distress your loved one experiences -- pain, trouble swallowing, choking, vomiting, and bathroom problems -- takes a toll on your own emotional well-being and can leave you feeling helpless and frightened. Depression often follows. Depression can rob you of initiative and the ability to make good decisions and compromise your caregiving ability. Help can be found for some routine care. Seek that help. Caregiving is not a one-person job. Don't think you can manage care by yourself. Seek out social support for yourself, as that is a buffer in times of stress. Exercise also helps prevent depression. Always remember that depression is an illness – getting depressed and getting help for it is not a sign of weakness.

To the reader:

Here we are in Massachusetts in the middle of summer. . long hours of daylight and beautiful sunsets, heat, humidity, and thunderstorms. My body moves better in heat than cold. Our family had a wonderful 6 day rafting trip on the Middle Fork of the Salmon River in Idaho earlier this month.

I hope you find helpful the new information in this issue of Synapse. You will notice that several of the articles are brief, giving computer links to journals and agencies but no content. I have three reasons for not printing entire articles: 1. Copyright laws, 2. Saving space and thereby printing costs, 3. All of us need to access sites such as the ALS Therapy Development Foundation, which are essential sources of ever-changing information.

If you cannot obtain a computer with Internet access at home, I suggest that you should contact your public library or ask a friend or family member to help you. Many business people are always upgrading and you might be able to find a computer for next to nothing.

I have mentioned in past issues how you can derive greater benefit from Synapse in the on-line version. You can click on all of the links and thereby reference a wealth of reference material, which is not readily available to you now. PLS Friends and PLS News are avenues to keep you connected to others with PLS who are dealing on a daily basis with the same issues you are. You are not alone. You may think you are too old to learn computers. Well, I'm in my 60s, not some young thing. I have bulbar problems and as my speech deteriorates, I find my computer my most effective means of communication. Please see what you can do, and let me know of any success.

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