Synapse

Spring 2005

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

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SP Foundation Announces National Convention and TeamWalk 2005

Columbus, OH September 30th, October 1st & October 2nd

- Assistive Technology, Adaptive Equipment & Kids
- Keynote Speaker: Dr. John Fink
  Leading researcher in HSP & PLS
- Friends, Food, Displays, Workshops
- Meet the Board of Directors, Open Q & A from the community to the Board
- TeamWalk And more…

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http://www.columbus.hyatt.com/
More details coming soon…
For more information contact Paul Brockman TEAM WALK Committee Chair
email to: paul@sp-foundation.org

New SPF Board

At the Annual Meeting of the Spastic Paraplegia Foundation held in Nashville, TN March 12-13, Annette Lockwood was named President. Mark Weber stepped down after serving for three years in that position. Mark said just before the vote that the SPF is facing new challenges that demand new leadership. Fortunately, he knew the perfect candidate for the job. He then nominated Annette Lockwood. Her confirmation was unanimous.
Also elected with unanimous votes were the following officers:
Vice-President: Linda Gentner
Treasurer: David Lewis
Secretary: Frank Davis
Re-elected to the Board were:
Mark Weber who will continue to serve as a Director, as Legal Counsel, and as chairman of the Research Committee
Paul Brockman
First term members elected:
Kris Brocchini
Jean Chambers, R.N.
Rick Pallas
Jim Sheorn

A Letter from our President

Dear Friends,
I am both excited and anxious about being the President of the Spastic Paraplegia Foundation. Excited because I have seen it grow from an idea to $500,000 in available grants for medical research in just three years. Anxious because now that we have come this far, we need to progress to the next level. Thanks to the Community and the Founding members of the Board, the Spastic Paraplegia Foundation is recognized as a viable organization. That is no small feat! But now we have decided to look at obtaining some professional nonprofit assistance to put together a Business Plan for SPF. The plan would include a Fundraising Plan with a goal of getting to the Million-Dollar level and sustaining that. Scientific research is expensive, according to Dr. Fink, expenses for laboratory mouse models of HSP were more than $100,000 last year alone. If you would like to help the Board of Directors put together this plan, please contact me at annette.lockwood@verizon.net.
On a final note, I want to welcome the new members to the Board - Kris Brocchini, Jean Chambers, Frank Davis (Secretary), Rick Pallas and Jim Sheorn. I also want to say a big Thanks to Kathi Geisler (former VP) and Marlene Doolen (former Secretary) for their endless support since the inception of SPF. Both of them have agreed to continue working with the Foundation, Kathi will continue publishing the E-News and keeping the website up-to-date and Marlene will continue to be the contact for handout materials at Connection meetings.
Continuing Board Members included Linda Gentner (VP), David Lewis (Treasurer), Paul Brockman and Mark Weber (former President).
We have our work cut out for us and appreciate everyone's support as we move forward in 2005.
Annette Lockwood, SP President

SPF announces 2004 PLS Research Grant Winner

The Spastic Paraplegia Foundation is pleased to announce the award of its' second 2004 Research Grant to Teepu Siddique of Northwestern University. Dr. Siddique's proposal is for a PLS Registry. The award is for $45,000 per year for two years. The PLS Registry will do several things. First it will collect and store blood specimen and other types of human cells from PLSers who agree to join the Registry. The PLS Registry will also collect medical and family histories on each person supplying specimens. Further, patients will be contacted on a periodic basis to determine if there are any changes in symptoms or diagnosis. Qualified researchers will be able to use the specimens and data to search for the cause of PLS. Teepu Siddique, M.D. is a Professor of Neurology, and Cell and Molecular Biology at Northwestern University Medical School in Chicago, Illinois. He is also the Director of the clinical Neuromuscular Disorders Program there.
The PLS Registry needs participation by every PLSer. The PLS Registry has been created to completely safeguard your privacy. To

accomplish this, two separate databases are kept, in separate computers, concerning the donors of each specimen. Only one database has identifying information about the person who supplied the specimen. Researchers have no access to that database or that computer. The other computer has info about the donors--but no identifying info--no names, address, SS's etc. Instead, each donor is assigned a number by the first computer. That number is the only thing about you contained in the second computer. And researchers are only given access to the second computer. They simply cannot determine who you are if they are working on your specimens. Further, the PLS Registry proposal was reviewed by the Institutional Review Board at Northwestern University. They insist that patient confidentiality is maximized. So, quite frankly, does the federal HIPPA law. No insurance company will ever have access to this data. The data is strictly, completely, private. To become a part of the registry please contact Nailah Siddique, RN, MSN, (Dr. Siddique's wife and colleague) at either (312) 503-2712 or email: nsiddique@northwestern.edu

Spastic Paraplegia Foundation Research Grant Program 2005

Contributed by Mark Weber

The Spastic Paraplegia Foundation (SPF) was created in 2002 as a voluntary, non-profit health organization. Our goals are:

1)to fund research to discover the cures for the upper motor neuron disorders hereditary spastic paraplegia (HSP) and primary lateral sclerosis (PLS), and

2)to provide educational materials, conferences and gatherings for people affected with and by these disorders, and

3)to foster support for those affected with and by these disorders, through both on-line and in-person contacts.

The SPF's ultimate goal is to see the day when all those with HSP and PLS are diagnosed, treated, and cured. With our research grant program we strive to make that day a reality.

2005 Research Grants

The SPF has budgeted $500,000 for its 2005 Research Grant Program. Grants in the range of $40,000 - $60,000 per year will be awarded for one and two year proposals. Proposals on any aspect of hereditary spastic paraplegia (HSP) or primary lateral sclerosis (PLS) will be supported. Research grants are offered primarily as "seed monies" to assist investigators with new ideas, those in the early or pilot phase of their studies, or as additional support for ongoing investigations with demonstration of need. We anticipate that studies funded by the SPF will develop into projects that can successfully attract future funding from other sources.

The title of each study funded by the SPF, the name of the principal investigator, institution, city and state will be published on our web page, as well as in our newsletter, annual report and wherever else the SPF feels is appropriate. Accordingly, each grant application must include a title understandable to the lay public that will be used as SPF wishes.

All other parts of the grant application are considered confidential and will only be released to members of the SPF Scientific Advisory Board, its consultants, and the Board of Directors.

Application Deadline: April 15, 2005

Awards will be announced by August 31, 2005.

Hand Crafted Quilt Made for SPF Raffle

The beautiful Falling Leaves quilt (shown on back page of Synapse) was made by Southern 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 coordinator is going to be new SP Board Member Rick Pallas. For now, please send a check made out to SPF for Rick at the address below.

Raffle tickets are: 1 for $5; 5 for $20; 10 for $30< Checks should be made payable to SPF and sent to:
Rick Pallas
4855 Lakeridge Street,
Apt. 2B
Ypsilanti, MI 48197

MEDICAL RESEARCH UPDATES

Research Snippets
Nature Biotechnology
http://www.nature.com/medicalresearch/index.html
Thousands of articles are compiled here. From this page you can query a disease, a type of cell such as motor neuron, or stem cell.

Antibiotics Protect Nerves in Mice by Turning on Genes, Large, Multi-Center Clinical Trial Planned in Lou Gehrig's Disease
A family of antibiotics that includes penicillin may help prevent nerve damage and death in a wide variety of neurological diseases, including Lou Gehrig's disease, dementia, stroke, and epilepsy. The antibiotics' beneficial effects, discovered in experiments in the lab and with mice, are unrelated to their ability to kill bacteria, the researchers report in the Jan. 6, 2005 issue of *Nature*. Instead, the drugs squelch the dangerous side of a brain chemical called glutamate by turning on at least one gene, thereby increasing the number of "highways," or transporters, that remove glutamate from nerves.
It would be extremely premature for patients to ask for or take antibiotics on their own," says the study's leader, Jeffrey Rothstein, M.D., Ph.D., director of the Robert Packard Center for ALS Research at Johns Hopkins and a professor of neurology and of neuroscience. "Only a clinical trial can prove whether one of these antibiotics can help and is safe if taken for a long time." A large, multi-center clinical trial planned for the spring will help determine the best dose of and schedule for ceftriaxone in people with ALS, and will measure whether the known risks of long-term antibiotic treatment are worth it, he says. The drug is currently approved by the U.S. Food and Drug Administration and used to treat bacterial infections in the brain.
"If we can find drugs that protect against other causes of nerve death in ALS, the combination might offer a real therapy, much like using drug combinations to treat cancer," says Rothstein. "The more we know about ALS and other neurological diseases, the better our chances of finding ways to prevent nerve death by all causes."
The research was funded by the National Institute of Neurological Disorders and Stroke, the Muscular Dystrophy Association and the Robert Packard Center for ALS Research at Johns Hopkins. The ALS mice were provided by Project ALS.

Botulinum Toxin Used in Children with Cerebral Palsy
SOURCE: Indian J Pediatr. 2004 Dec;71(12):1087-91.
http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=15630317&dopt=Abstract
Botulinum toxin is a neurotoxin that blocks the synaptic release of acetylcholine from cholinergic nerve terminals mainly at the neuromuscular junction, resulting in irreversible loss of motor end plates. It is being widely tried as a targeted antispasticity treatment in children with cerebral palsy. A number of studies have shown that it reduces spasticity and increases the range of motion and is particularly useful in cases with dynamic contractures. However improvement in function has not been convincingly demonstrated. It is an expensive mode of therapy and the injections need to be repeated after 3-6 months.

Whereas Botulinum toxin can be a valuable adjunct in select cases, it should not be projected as a panacea for children with spastic cerebral palsy.

Embryonic Stem Cells Can Become Motor Neurons

http://www.nature.com/dynasearch/app/dynasearch.taf

Researchers at the University of Wisconsin-Madison recently announced in Nature Biotechnology that they've developed a chemical recipe to coax human embryonic stem cells to develop into motor neurons, the muscle-controlling nerve cells that are lost in Xue-Jun Li and colleagues used two federally approved lines of stem cells and exposed them to retinoic acid early and to several other compounds later on. Vassilis Koliatsos, an MDA-supported neuropathologist studying stem cells at Johns Hopkins University, says the Wisconsin researchers' recipe works well in laboratory containers, but that putting them into animals will likely "hold many surprises." Koliatsos said it's unclear that the new motor neurons would survive in the face of an overwhelming degenerative process like that seen in ALS. And, he notes, even if they did survive, getting them to "talk to" muscle cells is another unmet challenge.

In ALS and other neurodegenerative diseases, Koliatsos says, every neuron lost increases the chance that more neurons will be lost. "It's theoretically possible that we can put in the right number of cells such that the neurodegenerative process can be delayed significantly," Koliatsos said, adding that not much else can yet be predicted.

Distinct novel mutations affecting the same base in the NIPA1 gene cause autosomal dominant hereditary spastic paraplegia in two Chinese families.

SOURCE: Hum Mutat. 2005 Feb;25(2):135-41.

http://www3.interscience.wiley.com/cgi-bin/abstract/109863905/ABSTRACT

Chen S, Song C, Guo H, Xu P, Huang W, Zhou Y, Sun J, Li CX, Du Y, Li X, Liu Z, Geng D, Maxwell PH, Zhang C, Wang Y.

Department of Medical Genetics, Zhongshan Medical College, Sun Yat-Sen University, Guangzhou, People's Republic of China.

Hereditary spastic paraplegia (HSP) is a neurodegenerative disease characterized by lower-limb spasticity, hyperreflexia, progressive spastic gait abnormalities, and an extensor-plantar response. It is genetically very heterogeneous, with 28 Human Genome Organization (HUGO)-approved IDs in the database (last search: August 8, 2004). Following the identification of the SPG6 gene, NIPA1, we have identified two novel mutations, c.316G>C and c.316G>A, in two independent Chinese families linked to the SPG6 locus. These two mutations would cause a p.G106R substitution, and cosegregated with the disease. Structural predictions suggest that p.G106 is located in the third transmembrane domain of the protein, and that the mutant p.G106R disrupts this structure, causing the intramembrane loop to descend into the cytoplasm. Our results identify two novel mutations responsible for HSP and suggest that c.316 of theNIPA1 gene may be a mutational hotspot.

Large deletion involving the 5'-UTR in the spastin gene caused mild phenotype of autosomal dominant HSP

SOURCE: Am J Med Genet A. 2005 Jan 6; [Epub ahead of print]

http://www3.interscience.wiley.com/cgi-bin/abstract/109863103/ABSTRACT

Iwanaga H, Tsujino A, Shirabe S, Eguchi H, Fukushima N, Niikawa N, Yoshiura KI, Eguchi K. First Department of Internal Medicine, Nagasaki University Graduate School of Biomedical Sciences, Sakamoto, Nagasaki, Japan.

Hereditary spastic paraplegia (HSP) due to mutations in the spastin gene (SPG4) located to 2p22-p21 is the most common form of autosomal dominant (AD) HSP. We performed PCR-based direct sequencing of SPG4, followed by a linkage analysis and subsequent Southern blot analysis in large Japanese kindred where 20 of 33 members were evaluated neurologically, and consequently 6 were affected with HSP. Clinical evaluation

showed that the mean age at disease onset of the patients was older and the disability was less severe than those of previously reported typical patients with SPG4 mutations. Direct sequencing of genomic DNA and RT-PCR product did not show a SPG4 mutation despite of a strong linkage to the SPG4 locus at 2p. Southern blot analysis suggested a deletion involving the 5'-UTR of SPG4. Further sequence analysis confirmed a heterozygous 2307-bp deletion spanning from the 5'-UTR to intron 1 of SPG4. The results suggested that transcription of the mutated allele starts from an authentic initiation site, but lacks an authentic translational start site of exon 1 because of a deficient splice donor site and coding region. The abnormal transcripts may result in rapid RNA decay. The novel refractory mutation we identified widens the spectrum of SPG4 mutations. These findings suggest that structural genomic abnormalities of SPG4 are more frequent than expected, and this explains previously reported cases more feasibly in which SPG4 mutations were failed to be identified but the disease was linked to 2p.

ALS & Professional Italian Football (Soccer) Players

Source: Brain. 2005 Jan 5; [Epub ahead of print]
http://brain.oupjournals.org/cgi/content/abstract/awh373v1

Chio A, Benzi G, Dossena M, Mutani R, Mora G. Divisione di Neurologia 2, Dipartimento di Neuroscienze, Universita di Torino, Torino, Italy.

Summary The cause of amyotrophic lateral sclerosis (ALS) is still unknown. A possible relationship between ALS and sport participation has been supposed, but never definitely demonstrated. We studied a cohort of 7325 male professional football players engaged by a football team from the Italian First or Second Division in the period 1970-2001. ALS cases were identified using different concurrent sources. Standardized morbidity ratios (SMRs) were calculated. During the years of follow-up, five ALS cases were identified (mean age of onset, 43.4 years). Three cases had a bulbar onset, significantly more than expected (P = 0.003). Since the number of expected cases was 0.77, the overall SMR was 6.5 [95% confidence interval (CI), 2.1-15.1]. The SMR was significantly increased for an ALS onset before 49 years, but not for older subjects. A significant increase of the SMR was found in the periods 1980-1989 and 1990-2001, whereas no ALS case was found in the1970-1979 period. A dose-response relationship between the duration of professional football activity and the risk of ALS was found (>5 years, 15.2, 95% CI, 3.1-44.4; </=5 years, 3.5, 95% CI, 0.4-12.7). Our findings seem to indicate that playing professional football is a strong risk factor for ALS.

Progress in Determining Cause of Juvenile PLS and HSP

Recessive motor neuron diseases: mutations in the ALS2 gene and molecular pathogenesis for the upper motor neurodegeneration
SOURCE: Rinsho Shinkeigaku. 2004 Nov;44(11):792-4.
http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=15651293&dopt=Abstract [Article in Japanese] Ikeda JE. Tokai University, Graduate School of Medicine, Neurodegenerative Diseases Research Centre.

We have initially identified a mutation in ALS2 as a causative for a juvenile autosomal recessive form of amyotrophic lateral sclerosis (ALS), termed ALS2 (OMIM 205100). ALS2 mutations also are causative for an autosomal recessive juvenile primary lateral sclerosis, and infantile-ascending hereditary spastic paralysis. To date, nine homozygous ALS2 mutaions from nine independent families have been identified. All of these mutations result in predicted premature translation termination caused by the recessive frameshift or nonsense mutation. ALS2 is a 184-kD protein comprising several putative guanine nucleotide exchange factor (GEF) domains [RLD; RCC1 like domain, DH. PH domain, VPS9; Vacuolar protein sorting 9 domain]. In vitro, ALS2 specifically binds to the small GTPase Rab5 and functions as a GEF for Rab5. Ectopic expression of full-length ALS2 has further

implied an association with endosomal membranes mediated by the VPS9 domain, consistent with ALS2 involvement in endosomal trafficking and fusion in conjunction with the activation of Rab5. These results combined with our findings suggest that an obstruction of endosomal dynamics might underlie neuronal dysfunction and degeneration in ALS2, PLSJ, and HSP, as well as in a number of other motor neuron diseases.

An RNAi strategy for treatment of ALS caused by mutant Cu,Zn superoxide dismutase gene.
SOURCE: J Neurochem. 2005 Jan;92(2):362-7.
http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=15663483&dopt=Abstract
Xia XG, Zhou H, Zhou S, Yu Y, Wu R, Xu Z.
Department of Biochemistry and Molecular Pharmacology, University of Massachusetts, Worcester, Massachusetts 01605, USA.
Amyotrophic lateral sclerosis (ALS or Lou Gehrig's disease) is a neurodegenerative disease characterized by motor neuron degeneration, paralysis and death. One cause of this disease is mutations in the Cu,Zn superoxide dismutase (SOD1) gene. As mutant SOD1 acquires a toxic property that kills motor neurons, by reducing the mutant protein the disease progression may be slowed or prevented. While mutant SOD1 is toxic, the wild-type SOD1 is indispensable for motor neuron health. Therefore, the ideal therapeutic strategy would be to inhibit selectively the mutant protein expression. Previously we have demonstrated that RNA interference (RNAi) can selectively inhibit some mutant SOD1 expression. However, more than 100 SOD1 mutants can cause ALS and all mutants cannot be inhibited selectively by RNAi. To overcome this obstacle, we have designed a replacement RNAi strategy. Using this strategy, all mutants and wild-type genes are inhibited by RNAi. The wild-type SOD1 function is then replaced by designed wild-type SOD1 genes that are resistant to the RNAi. Here we demonstrate the concept of this strategy.

United Kingdom Agency Grants Embryonic Stem Cell Research License to Study Motor Neuron Disease 09 Feb 2005
The Human Fertilisation and Embryology Authority (HFEA) has granted a license to the Roslin Institute in Edinburgh to create stem cells from embryos produced by cell nuclear replacement, a technique also referred to as therapeutic cloning. The license will allow researchers at the Institute to study Motor Neuron Disease, in particular, those patients whose condition cannot be linked to the genes already identified as causing the disease.
The Institute will use the technique to study stem cells made with the genetic material from patients with Motor Neuron Disease. Using these embryonic stem cells researchers can study the development of Motor Neuron Disease in patients who do not have the genes that are currently known to cause the disease. Whilst these embryonic stem cells would not be used to correct the disease, the study of these cells could help develop future treatments.
Angela McNab, Chief Executive of the HFEA said: "The HFEA's role is to ensure research on human embryos is only carried out when it is viewed as necessary under strictly defined guidelines, outlined in the Human Fertilisation and Embryology Act (1990). "We recognise that Motor Neuron Disease is a serious congenital condition Following careful review of the medical, scientific, legal and ethical aspects of this application, we felt it was appropriate to grant the Roslin Institute a one-year license for this research into the disease."
Notes to editors
The stem cells are made by combining skin cells donated by patients with Motor Neuron Disease whose condition cannot be linked to the genes already known to cause the disease, and eggs donated by women. The genetic material (nucleus) will be removed from the egg and replaced with the genetic material (nucleus) of the donated skin cell. The egg is activated and allowed to grow into a 5-6 day old embryo. Cells from the embryo will be taken out to develop

embryonic stem cells. Research on human embryos is only allowed for certain purposes. Under the initial Human Fertilisation and Embryology Act (1990) the HFEA could only grant licenses if it was satisfied the use of human embryos was for one of the following purposes:
-- To promote advances in the treatment of infertility
-- To increase knowledge about the causes of congenital disease
-- To increase knowledge about the causes of miscarriages
-- To develop more effective techniques of contraception
-- To develop methods for detecting the presence of gene or chromosome abnormalities
-- Increasing knowledge about the development of embryos
-- Increasing knowledge about serious disease
-- Enabling any such knowledge to be applied in developing treatments for serious disease
Human reproductive cloning is illegal in the UK. As a result of the Human Reproductive Cloning Act (2001) nobody in the UK is allowed to use cell nuclear replacement, or any other technique, to create a child.
The HFEA was set up in August 1991 as part of the Human Fertilisation and Embryology Act 1990. The HFEA's principal tasks are to license and monitor clinics that carry out in vitro fertilisation (IVF), donor insemination (DI) and human embryo research. The HFEA also regulates the storage of gametes (eggs and sperm) and embryos.
For further information please contact the HFEA press office http://www.hfea.gov.uk

First Generic Fentanyl Pain Patch Approved for Treating Severe Chronic Pain
Feb. 2, 2005 -- The FDA has approved the first generic version of the Duragesic Patch to treat people with severe chronic pain that cannot be managed with alternative pain killers. The generic version will be sold under the name Fentanyl Transdermal System. The approval will likely result in substantial savings for users of the pain management patch because generic versions of prescription drugs typically cost a fraction of their brand-name counterparts.
The agency granted Mylan technologies, Inc. approval to produce a generic version of Alza Corporation's Duragesic Patch. The pain patch is used to treat people with severe chronic pain that cannot be treated with alternative pain killers. When applied to the skin, the patch releases fentanyl, an opioid pain medication that is slowly absorbed into the body through the skin. Opioids are medications that fall within a class of drugs, which include morphine, codeine, and related drugs. These medications act to block the transmission of pain messages to the brain. The patch provides pain relief for up to three days. The original Duragesic Patch was approved in August 1990. It is currently approved to treat chronic pain in people who require continuous treatment with opioids and cannot be managed by acetaminophen-opioid combinations, nonsteroidal pain relievers, or short-acting opioids. Fentanyl is currently a schedule II controlled substance, which is the highest level of control for drugs used in medicine. Schedule II drugs come under the jurisdiction of the Drug Enforcement Agency (DEA) and are subject to manufacturing quotas set by the agency. The DEA considers medical need for a drug when establishing quotas. Schedule II drugs are also subject to distribution tracking, import and export controls, registration of prescribers and dispensers, and requirements for written prescriptions without refills.

Intrathecal Baclofen Therapy Over 10 Years.
SOURCE: J Neurosci Nurs. 2004 Dec;36(6):322-7.
http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=15673207&dopt=Abstract
Rawlins PK. Neuroscience Implant Program at Via Christi Regional Medical Center, Wichita, KS, USA. patrice_rawlins@via-christi.org
Intrathecal baclofen (ITB) therapy has evolved into a standard treatment for severe spasticity. After this therapy had been provided for 10 years, a retrospective chart review on 50 patients, representing a total 2,922 patient months of ITB

service, was done. These patients suffered severe spasticity from a number of disease processes including multiple sclerosis, cerebral palsy, and brain injury. The average dosage for the total group was 463 micrograms per day (microg/day), and 32% used a simple infusion mode. Pump refills occurred every 3 months for 58% of the group. Three evolving trends in ITB therapy were identified from clinical trial to current management: (a) higher catheter tip placement, (b) use of more complex infusion modes, and (c) a decreased complication rate.

Proneuron Announces Milestone Patent for Copolymer-1 (COP-1)

Patent expands use of Cop-1, previously approved for the treatment of MS

L.A., California, February 15, 2005 --- Proneuron Biotechnologies, www.proneuron.com, announced today the granting of U.S. Patent No. 6,844,314 for the use of Copolymer-1 (Cop -1) for protection from neuronal degeneration. The patent was granted to Proneuron Scientific Founder Professor Michal Schwartz of the Weizmann Institute of Science. The patent, exclusively licensed to Proneuron, is owned by Yeda Research and Development Company Ltd., the commercial arm of the Weizmann Institute of Science.

Cop-1 was invented at the Weizmann Institute of Science over thirty years ago and licensed to Teva Pharmaceutical Industries (NASDAQ: "TEVA") for the treatment of Multiple Sclerosis. Teva's drug, CopaxoneR, was approved by the FDA several years ago and has since been used safely and successfully. Years of research, led by Prof. Michal Schwartz of the Weizmann Institute of Science, has shown that Cop-1 acts as a low-affinity antigen that activates a wide range of self-reacting T cells, resulting in neuroprotective autoimmunity that is safe and effective against both CNS white matter and gray matter degeneration. The neuroprotective effect of Cop-1 vaccination was demonstrated in preclinical models of acute and chronic neurological injuries and disorders such as optic nerve injury, head trauma, glaucoma, Amyotrophic Lateral Sclerosis and Huntington's disease.

Under a collaboration agreement with Teva from 2001 and its 2003 expansion, Cop-1 is being developed by Teva as a therapy for several neurodegenerative conditions. Under this collaboration, a joint project is in progress to use Cop-1 for Huntington's disease, ALS and for the attenuation of the progressive optic nerve and retinal degeneration that causes visual field loss and eventually blindness in glaucoma patients. Cop-1 is expected to enter Phase II clinical trials in the near future for a number of indications.

"The evidence for the dual action of Cop-1, as an anti-inflammatory and a neuroprotective drug, has been fundamentally strengthened thanks to the research led by Proneuron and the Weizmann Institute of Science. Our collaboration with Proneuron presents us with a great opportunity to expand the use of this important drug," said Teva's V.P. of Strategic Business Planning and New Ventures, Dr. Aharon Schwartz.

For additional information, please contact Marjie Hadad, Media Liaison, Proneuron, Marjie.hadad@proneuron.com.

TROY: A Newly Identified Stop Signal in the Pathway for Nerve Regeneration

http://www.ninds.nih.gov/news_and_events/news_articles/news_article_regeneration_TROY.htm

One of the major puzzles in neuroscience is how to get nerves in the brain and spinal cord to regrow after injury. A new study has identified a protein, TROY, that inhibits nerve cell repair and plays a role in preventing nerve regeneration. This finding is an important step in developing new methods for treatment of spinal cord injury, stroke, and degenerative nerve disorders such as multiple sclerosis (MS).

Most of the cells in the human body have the ability to repair themselves after injury. However, neurons in the central nervous system (CNS) are unable to regenerate their injured axons. One of the major obstacles to regeneration in the adult CNS is the presence of inhibitory molecules that are associated with myelin, a fatty coating that forms a sheath around nerve cells. Research in recent years has focused on

identifying the chain of events that prevents regeneration.

Part of the inhibitory or "braking" machinery that stops nerve regeneration is Nogo, a protein normally found in myelin. Studies have suggested that a necessary partner or co-receptor to Nogo is a protein called p75, which is common in the developing nervous system but decreases during adulthood. However, while most neurons in the central nervous system do not have p75, these neurons still demonstrate myelin inhibition and can not repair themselves. In the new study, Dr. Zhigang He and colleagues from The Children's Hospital of Harvard Medical School in Boston asked, "Why do neurons without p75 still fail to regenerate after injury?" Their findings appear in the February 3, 2005, issue of Neuron. This research was funded in part by the National Institute of Neurological Disorders and Stroke (NINDS).

This study is the first to show that TROY is a critical player in blocking nerve regeneration. Although the research does not eliminate the possibility that other molecules are also involved in regeneration, the study helps scientists to understand how myelin inhibition may be regulated. The finding that more than one protein may be involved in myelin inhibition adds a new level of complexity to designing therapeutic strategies for treating CNS injury.

The discovery of the TROY protein enhances understanding of nerve injury and provides a piece in the puzzle of CNS nerve regeneration. Investigators hope this information will point to ways to stimulate nerve repair in the brain, which is vital for restoring functions in persons with MS, spinal cord injury, stroke and other CNS injury conditions.

Reference: Park JB, Yiu G, Kaneko S, Wang J, Chang J, He Z. "A TNF receptor family member, TROY, is a coreceptor with Nogo receptor in mediating the inhibitory activity of myelin inhibitors." Neuron, February 3, 2005, Vol.45, pp.345-351.

--By Michelle D. Jones-London, Ph.D.

Deciphering the Genetic Babel of Brain Cell

Ed. Note: Drs. Molyneaux and Arlotta were two of our speakers at the SP Foundation National Convention in 2004

Two major questions in the fields of neural development and neural stem cell biology are what genes and molecules control the specific development of the many distinct types of brain neurons from stem cells, and how these genes and molecules might be manipulated to control stem cells toward replacement of specific types of damaged brain neurons. Now, researchers led by Jeffrey Macklis, Bradley Molyneaux, and Paola Arlotta of the MGH-HMS Center for Nervous System Repair at Massachusetts General Hospital and Harvard Medical School, and the Harvard Stem Cell Institute, have identified major elements of such a program of control genes for corticospinal motor neurons (CSMN), critical neurons that connect the brain to the spinal cord, subject to degeneration and injury in multiple human neurological diseases.

In their report, the scientists isolated corticospinal motor neurons (CSMN) at several stages of their development, and analyzed the genes that are active in these neurons during their progressive development in mice. They compared these CSMN genes with those of two other closely related subtypes of cortical neurons to discover specific genes that are seemingly critical to CSMN development. They then confirmed the central role of these genes by genetic deletion of one identified gene, resulting in total elimination of connections from the cerebral cortex to spinal cord. CSMN, which connect the cortex and spinal cord, carry the signals that control movement, or motor function. These neurons are important because they are the brain neurons that degenerate in amyotrophic lateral sclerosis (Lou Gehrig's disease) and their damage contributes critically to the loss of motor function in spinal cord injury. Better understanding of the genes that control the development of these neurons could aid in the development of treatments for these disorders. For example, some of these genes and molecules could be manipulated to enhance the survival or

function of diseased corticospinal motor neurons, or these signals might be used to control stem cells to replace diseased CSMN and rebuild functional connections.
The entire article may be found in New hope: 'Conditional approval' for ALS study
Dr. James Bennett of UVA is seeking approval to resume trials with the drug, pramipexole, a chemical mirror image of a drug already on the market to treat Parkinson's Disease. In 2004 he treated 40 ALS patients with pramipexole. Remarkable results were seen with improved use of hands and speech. The FDA has given him the go-ahead for compassionate use. He now has theconditional approval of UVA but must raise funds both for the drug and a project coordinator. More information about the program may be found in March 10, 2005 in issue 0410 of The Hook
http://readthehook.com/stories/2005/03/10/newsNewHopeconditionalAppr.html

Progress on Familial ALS
http://www.pnas.org/cgi/content/abstract/0408277102v1
Small-molecule-mediated stabilization of familial amyotrophic lateral sclerosis-linked superoxide dismutase mutants against unfolding and aggregation.
Ray SS, Nowak RJ, Brown RH Jr, Lansbury PT Jr.
Harvard Center for Neurodegeneration and Repair and Department of Neurology, Harvard Medical School, Boston, MA 02115; Center for Neurologic Diseases, Brigham and Women's Hospital, 65 Landsdowne Street, Cambridge, MA 02139.
Familial amyotrophic lateral sclerosis (FALS) is a fatal motor neuron disease that is caused by mutations in the gene encoding superoxide dismutase-type 1 (SOD1). The affected regions of the FALS brain are characterized by aggregated SOD1, and the mutations that destabilize SOD1 appear to promote its aggregation in vitro. Because dissociation of the native SOD1 dimer is required for its in vitro aggregation, we initiated an in silico screening program to find drug-like molecules that would stabilize the SOD1 dimer. A potential binding site for such molecules at the SOD1 dimer interface was identified, and its importance was validated by mutagenesis. About 1.5 million molecules from commercial databases were docked at the dimer interface. Of the 100 molecules with the highest predicted binding affinity, 15 significantly inhibited in vitro aggregation and denaturation of A4V, a FALS-linked variant of SOD1. In the presence of several of these molecules, A4V and other FALS-linked SOD1 mutants such as G93A and G85R behaved similarly to wild-type SOD1, suggesting that these compounds could be leads toward effective therapeutics against FALS.
Source: Proc Natl Acad Sci U S A. 2005 Feb 28; (Epub ahead of print)

Baclofen Overdose
http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=15730591&dopt=Abstract
An unusual cause of overdose after baclofen pump implantation: case report.
Psaty EL, Abbott R.Division of Pediatric Neurosurgery, Beth Israel Medical Center/Institute of Neurology and Neurosurgery, New York, New York. OBJECTIVE AND IMPORTANCE: Intrathecal baclofen delivery for the treatment of spasticity has been used for almost 20 years with a great deal of success. A wide variety of complications and pitfalls have been described. This report details a novel complication involving inadvertent and initially unrecognized canalization of the subdural space with the spinal catheter, which ultimately resulted in an overdose. CLINICAL PRESENTATION: An intrathecal pump system was implanted in a 15-year-old girl with spasticity. This initially resulted in a lack of therapeutic effect. The diagnostic workup ultimately led to contrast administration through the pump system, which precipitated a baclofen overdose when sequestered medication in the subdural compartment was released into the intrathecal space. INTERVENTION: The spinal catheter was subsequently revised, and the patient made a full

recovery. CONCLUSION: The possibility of a subdural catheter should be included in the differential diagnosis in patients who experience a lack of drug effect after pump implantation, despite increases in dosage. Close monitoring is required because of the risk of spontaneous or induced overdose, which may occur when a communication develops between the subdural and intrathecal compartments.
SOURCE: Neurosurgery. 2005 Mar;56(3):624.

Assessing Driving Capability: A method for individual testing
The significance of paraparesis inferior studied in a controlled experiment.
SOURCE: Appl Ergon. 1991 Apr;22(2):75-84.
http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=15676801&dopt=Abstract
Lings S. Chief Physician, Department of Occupational Medicine, Odense University Hospital, DK-5000 Odense C, Denmark.
The part played in traffic safety by driver illness or disability is uncertain or unknown. So also are the specific identity and degree of the disorders which necessitate the use of driving aids or which completely incapacitate a person from driving. Despite the gravity of the problems, the question of fitness to hold a driving license is decided throughout the world mainly on the basis of subjective assessment. Controlled experiments exploring the significance of disorders have only been carried out on a restricted scale. In this paper a description is given of a mock car, which is used both for research and individual assessment. It enables the measurement of strength application, steering wheel turn speed, simple reaction times when operating pedals and steering wheel, erroneous reactions, and choice reaction times. Experiments involving 109 able-bodied and healthy persons showed, as expected, that the muscular strength of men was greater than of women, and that men were significantly quicker at carrying out functions which primarily depend upon speed of movement and of strength. Apart from this, however, there were no significant sex-related differences. Almost all variables showed age dependence, this being most pronounced in the case of men. Thirty-two percent of the test candidates committed errors like braking instead of turning the wheel or turning to the wrong side. Neither the incidence nor the seriousness of errors bore any relation to sex or age. Fifty-two persons suffering from paraparesis inferior were compared with the 109 able-bodied subjects. The degree of paresis co-varied with reaction times, but the degree of spasticity only to a minor extent. The results indicate that at a speed of 80 km/h, 'slight paresis' increases reaction distance by around 2-3 m (15%), and 'moderate paresis' by the region of 50 m.

Webcast Neurodegenerative Disease Markets: Opportunities to Solve the Mysteries of Alzheimer's, Parkinson's, ALS, and Other Devastating Illnesses
Contributed by Lynn Holmes
A unique look at the common threads that link ALS and other neurodegenerative diseases is explored in a webcast that focuses on the research underway by many biotechnology companies to provide new answers and pathways to treatments and cures. Dr. Cathy Lomen-Hoerth, Director of UC SF Medical Ctr. ALS Patient Care & Research Center and ALS Assn. Science Director Lucie Bruijn is featured on the ALS webcast, produced by Killen & Associates, a highly-respected research firm based in Palo Alto, CA. The Killen webcasts look at the possibility of following a shorter path to finding cures for ALS and other neurodegenerative diseases by opening up a multi-billion dollar marketplace for innovative biotech and pharmaceutical companies. Firms that make breakthroughs in one disease area may have the technology necessary to open doors to other neurodegenerative disease markets as well. The experts' insight creates a foundation for envisioning how biotech and pharmaceutical firms can shorten the time to find cures by leveraging the common disease elements. To view a multimedia trailer of the series, visit http://www.killen.com/ndd/trailer. The series is available for general viewing at

http://www.killen.com/ndd. This webcast will be available for 18 months at this site.

Oral Analgesics for Acute Nonspecific Pain
Dolores Carron suggested including this information from the American Association of Family Physicans.
Physicians most often recommend or prescribe oral medication for relief of acute pain. This review of the available evidence supports the use of acetaminophen in doses up to 1,000 mg as the initial choice for mild to moderate acute pain. In some cases, modest improvements in analgesic efficacy can be achieved by adding or changing to a nonsteroidal anti-inflammatory drug (NSAID). The safest NSAID is ibuprofen in doses of 400 mg. Higher doses may offer somewhat greater analgesia but with more adverse effects. Other NSAIDs have failed to demonstrate consistently greater efficacy or safety than ibuprofen. Although they may be more expensive, these alternatives may be chosen for their more convenient dosing. Cyclooxygenase-2 inhibitors provide equivalent efficacy to traditional NSAIDs but lack a demonstrable safety advantage for the treatment of acute pain. For more severe acute pain, the evidence supports the addition of oral narcotic medications such as hydrocodone, morphine, or oxycodone. Specific oral analgesics that have shown poor efficacy and side effects include codeine, propoxyphene, and tramadol.
To read the entire article, push down control while you click:
http://www.aafp.org/afp/20050301/913.html

LIVING WITH PLS/HSP

A Request for PLS Information
from Carolyn Sartain Anderson
Friends,
My youngest daughter, Margaret, is a Psychology major at George Mason University in Fairfax, VA. She has chosen the topic, 'Coping with Primary Lateral Sclerosis: Patient and Family' for a mega-paper.
Although I am also enlisting the aid of my friends who have been diagnosed with HSP, I need your extremely valuable input also, as we are in this together. Please e-mail or snail-mail me all responses asap and I will forward them to Margaret. Any format is acceptable; make your response as lengthy/brief depending on your time/energy level. Margaret is hoping for some nice, long descriptions!
1. Year/age diagnosed
2. Age now/state where you reside
3. Where you are in progression (consider walking/speech/working/eating/dietary considerations/you name it)
4. Where diagnosed/how Dr. shared/how you felt and your family (I realize this is very personal info) upon learning about diagnosis
5. How you learned more about our illnesses
6. How you cope physically and emotionally
7. How your family is coping with your illness emotionally and supporting you

Your names will not be used, unless you give me permission; instead may Margaret identify you by age and state?
I am in hopes that Margaret's paper may be featured on the SPF website. I am so very proud of her for championing our cause and enlightening others as to 'who we are'.
Please e-mail: beachmusic1012@bellsouth.net
Street address: 262 Fig Street,
Sebastian, FL 32958
Many thanks in advance for your help on Margaret's paper.

HSP Support Group in Australia
Contributed by Mark Weber
I have heard that there is an HSP support group in Sydney, Australia. It will be run, I think, by Dr. Garth Nicholson of Concord Hospital at the University of Sydney. For more details please contact Dr. Nicholson's office at 61-2-9767-6202. This may be limited to those with HSP only.

Cruises
Contributed by Michael A Breaux < mabreaux@earthlink.net>
I have been on many Cruises in a wheelchair, and

loved all. Be aware the some of the shore trips will not be accessible to a wheelchair, but there is always something to do. I had my service dog along with me I had his company and we were great. Quick note: my service dog was allow on the ship, but was not allowed in port in most foreign ports so he slept in my stateroom while we were gone. Great experience, loved every one of them.

Grab Bars

Contributed by Dolores Carron

I'm in the process of having my bathroom modified for a "no ledge" shower. I found a wonderful on-line site for reasonably priced, excellent quality grab bars, shower seat, and related items. They shipped in just 3 days. Very complete installation instructions were included. I liked them because they offered grab bars, etc. that were white, rather than the stainless steel ones. I'd recommend them to anyone in need of such items. Their on-line site is www.grabbarspecialists.com or 1-800-707-5033.

Living Will, Health Care Proxy, DNR

Contributed by Editor

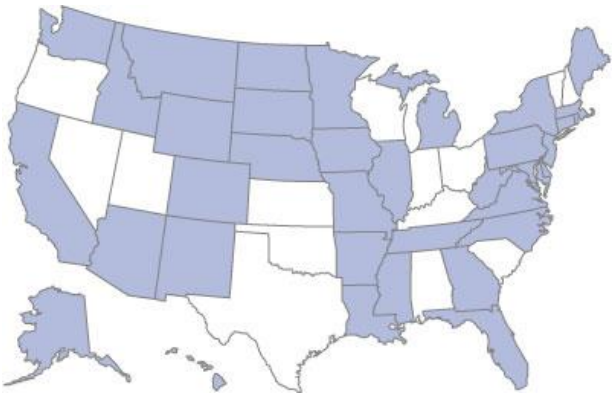
This spring everyone in the US has been made aware of the advisability of having written documents in place which specify how you would like to be cared for at the end of your life as well as during any event such as surgery, in which you are temporarily not able to make your own decisions. According to the media, only 20% of adults have Living Wills. I thought it would be a valuable service to our patient community to provide you with the link to a reliable resource on Living Wills, Health Care Proxy and Do Not Resuscitate matters.

An attorney friend suggested Aging with Dignity, http://www.agingwithdignity.org/ The copyright booklet they publish, Five Wishes, is comprehensive.

Below, I have listed the five wishes covered in the booklet:

1.Which person you want to make health care decisions for you when you can't make them.
2. The kind of medical treatment you want or don't want.
3. How comfortable you want to be.
4. How you want people to treat you.
5.What you want your loved ones to know.

The twelve page booklet has forms to fill out and options to cross out if you do not want a particular treatment. Each person for whom you need a living will need their own copy of Five Wishes. Below is a map showing 35 shaded states where the Five Wishes document is acceptable.



For those without computers call: 1-888-5-WISHES (594-7437)

I found reading the many options was in and of itself a valuable learning exercise. Aging with Dignity and many media outlets are stressing the vital importance of families discussing end of life matters now, before critical decisions must be made.

Communication Aids

Contributed by Don Wilson

don-wilson@earthlink.net

One of the greatest benefits of our Internet groups is the relative ease of communication. Even though many work long and hard typing a message, sometimes with only one finger, their thoughts can be communicated to others.

TDD relay allows those with impaired hearing communicate. I speak to the Relay Operator who then types what I am saying into a terminal which sends it to the hearing impaired person's TDD machine where he/she reads my comments. To let the operator know when each has finished talking (and need to take a breath), they then say "Go ahead" which tells the operator that you have

stopped speaking.
But what about telecommunication solutions for people with speech disabilities? There is something new to assist those with such disabilities. It is called SPEECH-TO-SPEECH (STS). Operators are being trained (some have been already) to understand mild disabilities and to understand synthetic speech generated by a piece of speech augmentation equipment, such a a Link, Pollyanna or Enkidu.If using this service with speech, a number is dialed and an operator will ask "May I have the number you are calling?" The caller gives the area code and number, name of the person or company being called, and any special instructions, such as how to repeat what is being said or leaving a message on answering machine). When the connection is made, the operator will say "Go ahead" as the cue for the caller to begin speaking. The operator will repeat what is said as needed, or could ask that the caller repeat a word or parts of the message. The caller needs to say "Go ahead" when it is time for the operator to begin speaking.
To make a call using communication devices, the same steps are followed in dialing. It is suggested that a speaker phone be used, placing the communication device in front of the speaker phone. The caller would have already set up a phone call page with pre-programmed short phrases or words. One might be: "I am using a communication device. Please wait until I make my selections. Another would be "yes" or "no". "My name is --------and I would like to speak to ----" would also be helpful as a pre-programed message. Others could be "Thank you", "Please wait", "Please repeat what I said".
There are some tips for success:

1. Consider alternative strategies for dialling
2. Post the number for STS on the phone
3. Program the STS number in the phone
4. Adjust the volume on the phone
5. Allow extra time for making calls
6. Relay operators may be busy, call again
7. Trial and error process to learn the system
8. Expect silence and wait time during the call.

We saw this new system in use at the Assistive Technology EXPO last November. During the presentation, the instructor used the system with a Light Writer to order a pizza. It arrived just as the session ended and was actually lunch for the presenter.
This service is part of the Relay service for each state and is supposed to already be in use. There may or may not be a special number to put a caller in direct touch with a relay operator as by calling 711, where you will generally get a TDD operator, who then
should be able to switch you to a STS operator or give another number.
This could be a dramatic improvement in communications for those with speech disabilities. Check it out in your state.
Ed. Addendum
Here is the US government Federal Communications Commission site on Speech to Speech: http://ftp.fcc.gov/cgb/dro/sts.html
Open this link, http://www22.verizon.com/foryourhome/SAS/TelecomRelayServices.asp and you'll learn what is available in your state from Verizon.

Websites to Better Understand Spasticity
http://www.wemove.org/spa/spa.html
http://www.exploringspasticity.com

The TAO of DI #26
dianamj100@hotmail.com
This a copy of a monthly thing I write and send to all my friends. It's called The TAO of DI. Just little bits of my experience and my weird slant on life....would you like to get one each month?
Well, I am finally getting used to my new power wheelchair. I say, "finally", because it isn't as easy as it looks! In the first place....I have cats and dogs. Those of you who have pets will automatically understand. They are curious creatures....especially the cats. After one or two initial 'shrieks' and 'hisses', I have learned to avoid errant tails and paws. Then, of course, there is my Labrador-Retriever dog who, unfortunately, is friendly, curious, has a sense of humor and a nose that is roughly on the same level as mine when I am sitting down....figure that one out for

yourselves! Suffice to say it made for a few startling moments!
I have also learned that if you stop to reach for something, briefly turn the power off. A loose sleeve can easily catch on the power toggle. This I discovered after, not one, but two episodes of crashing my forehead into the kitchen counter (add appropriate bad language here). In addition to all of this, I have learned that there are several categories
of people, in terms of 'reactions' to someone in a wheelchair. Let's take shopping as an example:
1. Those who politely open doors and help me reconstruct neatly stacked store displays.
2. Those who, for some reason, assume I am deaf and articulate their words in a slow, loud voice (my personal favorites!).
3. Those who walk past, notice the chair and automatically say "I'm sorry" even though
they are at least 3 feet away ("I'm sorry too...but you can still walk past me!").
4. Those who continuously walk through stores with their heads down, often resulting
in a close but friendly encounter in my lap.
5. Those who like to walk backward while saying goodbye to someone, causing me to hit reverse with startling and deadly speed!
6. Those who actually make eye contact and give me a warm smile as they pass.
Number 6 is actually me favorite category. One of the blessings of going shopping in a wheelchair is that you often have more warm and personal contact with people. It can lift my mood way up into the stratosphere! Without the chair, most people don't me at all! I suppose you can look at such a situation from 2 viewpoints. You could feel embarrassment, resentment and sadness or you could experience humor, a sense of fun and a gloating realization that you get to sit and relax while everyone else has to huff and puff around miles of department store!
I guess many of life's unexpected and unwanted situations can be viewed from both of these perspectives. It is just a matter of making a conscious effort to chose which one. If the effort fails.....try again.....and again. Thanks again, my dear friends, for all of your kindness, and support! Bonus! I love you all. Di

Tax Deductions for the Disabled
by Christina Medvescek for the MDA
"Anything you had to buy that your next-door neighbor didn't have to buy is probably a tax deduction."
That's the basic test that Armand Legault, a former auditor for the Connecticut Department of Revenue Services, advises taxpayers with neuromuscular diseases to apply to the question: "Is this deductible?"
Did you need extra electricity to run a BiPAP at night in 2003? Have to pay extra for an accessible van? Did you have to remodel your bathroom or build a ramp? Send your child to a special school? Hire someone to help you get dressed in the morning? Write it all off.
"If you had no choice in the matter, that's the key," says Legault, who spent 33 years as a state tax auditor, and who now gives tax seminars for people with disabilities, certified public accountants (CPAs) and other tax preparers. Legault, of Newington, Conn., has spinal muscular atrophy and was MDA's 1992 Personal Achievement Award winner for Connecticut.
Are most people with disabilities aware of the tax breaks they can claim?
"Oh gawd no!" laughs Legault. For example, he says, many working people with disabilities have no idea of the wealth of deductions available to them as "impairment-related work expenses." (See instructions in IRS Publication 502, listed in "Tax Help Resources.")
'A Gold Mine'
As an incentive to keep people with disabilities working, the federal government allows full- and part-time workers to deduct unreimbursed business expenses "that are necessary for you to work." To qualify, you must have an impairment "that substantially limits one or more of your major life activities, such as performing manual tasks, walking, speaking, breathing, learning and working."
These business deductions are more valuable than medical deductions because every penny is

deductible, whereas medical expenses must exceed 7.5 percent of your adjusted gross income before they can be deducted. (Adjusted gross income, or AGI, is the total of your income, minus a few adjustments. It's the figure listed on line 34 of your 1040 form.)

In addition, unlike standard business expenses, disability-related business expenses aren't capped at 2 percent of AGI. This means, says Legault, "If it comes out of your pocket and it is required in order for you to be an employable person, you can deduct it as a nonreimbursed employee expense. "It's a gold mine for people who are working." These deductible expenses include:

- Home accessibility remodeling required for work, such as a roll-in shower or a lowered kitchen sink (because you can't go to work dirty or hungry), or a ramp (because you can't go to work if you can't leave the house) The cost of a personal care attendant who helps you get ready for work or helps you at work
- The unreimbursed purchase, repair and maintenance of equipment necesary to get you up, out of the house and working productively, such as a lift, wheelchair, shower chair or BiPAP
- The extra cost of electricity required by such equipment
- The food, grooming and medical care costs of a service animal
- Specially adapted work equipment you buy yourself, like a special computer keyboard
- Meals and extra expenses for a live-in attendant. Although your meals aren't deductible (your next-door neighbor has to buy food, too), if your live-in assistant helps you get ready for work in the morning and you provide breakfast as part of his or her compensation, the cost of that food is a business expense. Additional utility expenses also may be considered under this category. Legault had this arrangement with three live-in assistants before he retired.

Transportation costs can be a huge tax-saving bonanza for many disabled employees, amounting to several thousands of dollars in deductions. For example, if you drive a modified or accessible vehicle, you may deduct your daily mileage to and from work (currently 36 cents a mile, compared to 12 cents a mile for medical mileage). You must be able to show that you can't use some other form of transportation, like a regular car, bus or carpool, due to your disability (or because they're not available).

(Another little-known fact: About 70 percent of states offer a break either on local property tax or vehicle license fees for accessible and modified vehicles, Legault notes.)

Even if you don't drive, you can deduct the cost of hiring special transportation to get to work, such as an accessible van service.

You could even buy your own van and hire someone to drive you to and from work, and deduct the extra cost of the accessible vehicle, mileage and the cost of the driver, Legault says.

Gray Area

Legault warns, "Most CPAs and tax preparers are unaware of this whole thing. They only think in terms of medical deductions. No! This is better." One reason for the confusion is that impairment-related work expenses are a vague, gray area in the tax code. Legault, who supervised tax auditors for years, recommends making good use of the absence of strict definitions.

"If you can prove that you need it to work — that's the key. Your doctor says you need it or you can show you can't work without it — then take it [as a deduction]!"

He notes that, in the absence of clear definitions, IRS auditors aren't on solid ground either. If you believe strongly that you deserve the deduction and aren't taking advantage of the system, "argue that you're an employee and you need this to work," he advises. "You'll win."

Medical Deductions

If you're unemployed, or if the person with neuromuscular disease is your child, spouse or dependent, then you're looking for medical rather than business deductions.

You may deduct out-of-pocket, unreimbursed medical expenses that exceed 7.5 percent of your adjusted gross income. For example, if your AGI is $30,000, and you had $5,000 in medical expenses, you may deduct expenses over $2,250

(7.5 percent of AGI), leaving you $2,750 in deductions. These expenses must relate to the "diagnosis, cure, mitigation, treatment or prevention of disease."

Attendant Care Expenses: Credit or Deduction?

If attendant care costs are for a child, spouse or other nonemployed dependent, taxpayers have two choices: Claim the cost as a medical deduction or take a child/dependent care credit (see IRS Publication 503).

A *deduction* lowers the amount of income on which you're taxed, indirectly lowering your taxes. A *credit* is a direct dollar-for-dollar reduction in tax — e.g., a $100 credit lowers your tax $100.

You can claim attendant care expenses as either a credit or a medical deduction, but not both. (However, if you choose the credit, any attendant care costs in excess of the credit's limits may be claimed as medical deductions.)

To qualify for the child/dependent care credit, the care recipient must be a child under 13 or someone unable to either physically or mentally care for himself without attendant care. In addition, the care must be provided so that the taxpayer(s) can work or look for work. The credit ranges from 20 percent to 35 percent of expenses, up to $3,000 a year ($6,000 for two or more qualifying people).

In general terms, a credit is better than a medical deduction. But because tax savings vary depending on a variety of factors, especially the income of the taxpayer, do the math to see which approach would save you more.

Back It Up

Keep all relevant prescriptions, doctors' letters, receipts, credit card statements and cancelled checks. Make sure prescriptions specify that the expense is to mitigate the effects of your neuromuscular disease, not just for general health. If you're claiming the cost of an item such as extra electricity or food, be sure to have receipts and records that show the price discrepancy. You don't need to turn these in to the IRS, but should keep them on file in case you're challenged.

If you missed taking a deduction or credit in a previous year, you can file an amended tax return for up to three years after the purchase, using IRS Form 1040X.

Explore all your tax options, Legault recommends. Don't assume professional tax preparers are aware of all deductions available to you; carefully outline the specifics of your unique situation for them — and don't be afraid to do your own taxes.

Not every taxpayer affected by a neuromuscular disease can benefit from tax deductions, but it's a shame to miss out if you're eligible, he says. "It's due you. Why should you have to buy this expensive van or wheelchair that your neighbor doesn't have to buy? Fight for your cause!"

IRS Publications

These publications are available online at www.irs.gov or by calling (800) 829-3676.

Medical and Dental Expenses, Publication 502. Outlines what is and isn't deductible as a medical expense or impairment-related work expense.

Tax Highlights for Persons with Disabilities, Publication 907. Gives a brief introduction to tax laws of interest to people with disabilities and caregivers.

Child and Dependent Care Expenses, Publication 503. Tells who qualifies and how to figure and claim this credit.

Credit for the Elderly or the Disabled, Publication 524. Outlines who is eligible for this credit, and how to determine and claim it.

Internal Revenue Service Programs

(800) 829-1040

Tax Counseling for the Elderly (TCE) and Volunteer Income Tax Assistance (VITA) provide free federal, state and local income tax assistance for lower-income, elderly and disabled taxpayers.

Taxpayers with incomes under $60,000 may take advantage of the service, provided their returns are "simple." Volunteers are trained to prepare basic forms and forms claiming credits such as the earned-income credit and the child credit.

AARP Tax-Aide

(888)227-7669
www.aarp.org/taxaide
This free program operates under a cooperative agreement with the IRS. It's available to help prepare tax returns and answer questions for people of all ages with middle and low incomes, with a special emphasis on those over 60. Volunteers are trained and certified by the IRS and are stationed at more than 10,000 sites around the country. Home visits also are available. The program is available from Feb. 1 through April 15.

CAREGIVING

Canine Caregiving

Contributed by Thurza Campbell

I'd thought for a long time that our Black Lab, Euro, might be trainable to become a service dog. Often when I need to pick something up, or do a down on the floor task, or when I fall I need help getting up. Euro is almost three, so when we contacted Janet Harris an excellent dog trainer, she was doubtful that he could be taught to "stand" while I'd tug and push. Another complicating factor is that I can't talk, so he must respond to hand commands only. The one private lesson was a smashing success, as the pictures on the back page of Synapse attest. The key to success is that he wears a harness for "work". Harness means he's on duty. We take it off for play time. Initially Jim did the daily training, but now I manage fine.

Well Spouse

Illness and accident attack without warning and can happen to anyone. No two people are living in the same situation and no two illnesses exact the same toll. Alzheimer's is different from MS, heart disease is different from Parkinson's, stroke is different from accident. But all well spouses face similar problems of anger, guilt, fear, isolation, grief, and financial threat whether they are full-time caregivers or whether their partners have only moderately disabling illnesses.

Well Spouse is a national, not for profit membership organization which gives support to wives, husbands, and partners of the chronically ill and/or disabled. Well Spouse support groups meet monthly. Here, our members can share their thoughts and feelings openly with others facing similar circumstances in a supportive, non-judgmental environment. WS support groups are also an excellent source for information on a wide-range of practical issues facing spousal caregivers. Well Spouse support groups exist or are being formed in many areas of the country. To contact a support group near you or to learn how to start a group, please go to our "SUPPORT GROUPS" page.

http://www.wellspouse.org/

Mailing address: 63 West Main Street -- Suite H, Freehold, NJ 07728
Phone number: 1-800-838-0879
Office number: 1-732-577-8899
FAX number: 1-732-577-8644

Creative Use of Walkers

Look who's riding in Annette's walker

SP Foundation Board

l-r: Mark Weber, Eddie Adcock, Jim Sheorn, Rick Pallas, Jean Chambers

l-r: Kris Brocchini's sister-in-law, Kris, Linda Gentner, David Lewis, Craig Gentner, camera shy person, Jeanne Kroll

Ontario, Canada Quilters

The ladies from South Ontario, who crafted the Turning Leaves quilt

Turning Leaves Quilt

Quilt's underside with inscription.

Norman, Oklahoma SP Connection

l-r: Bill Wetzold, Eric Talla, David Canan, Vanessa Reyes, Lillian Wetzold, and Mark Dvorak. Lynn Parli, Margaret Stribling, Dee Stribling, Marvin (Marv) Wiebener, Peggy Wiebener, and Anne Marie McLennan .

Canine Caregiving

Editor pulls herself to knees while leaning on Euro

Now pushing hard on Euro she is able to stand upright