



MOUNT SINAI
SCHOOL OF
MEDICINE

Spring 2004

The MDA/ALS Beacon

The Muscular Dystrophy Association/Amyotrophic Lateral Sclerosis
The Mount Sinai Medical Center



Word from the Director

With great pride, I wish to introduce The MDA/ALS Beacon. This semi-annual publication is a labor of love and dedication on the part of our spectacular team of professionals, who are committed to caring for patients and their families with Amyotrophic Lateral Sclerosis and related diseases. Every person and family afflicted by ALS faces a huge range of practical and emotional challenges, from transportation to managing families, from hope to despair. Our hope is that The MDA/ALS Beacon will create an interactive forum to discuss problem-solving skills that make it easier to cope with all these challenges. Toward that end, we are thrilled to have Meryl Houghton join us as our editor on matters of giving care to loved ones with ALS in the *On Caregiving* column. Dorothy Kolb will be our editor for patient issues. Members of our team will also contribute helpful tips that they have discovered during their years of service to our patients, and we will keep you informed of advances in research. A special treat in this issue is an essay from Eileen Kellner, the founding administrator for the Mount Sinai ALS program who in 1976 created the very first multidisciplinary program for ALS in the country. I would also like to thank the Muscular Dystrophy Association for their unyielding support for our clinical and research programs. Special thanks to Nancy Xenakis, CSW, MS and Lina Mina, RN, EdM for their tireless work and dedication to our patients with ALS. They are truly very special people and we are incredibly fortunate to have them both with us.

To all whose life has been touched by ALS, we bring The MDA/ALS Beacon. We hope that we will help each other as we work toward the cure we so desperately want.

Dale J. Lange, M.D.

Director, Division of Neuromuscular Disease

Research Update In Brief

Funding and conducting research is critical in our quest to find a cure for ALS and in each issue we will bring you updates from the research perspective. In April, Dr. Giulio Pasinetti, MD, PhD, Professor of Neurology at Mount Sinai School of Medicine, presented his findings describing an ALS marker at the American Academy of Neurology Annual meeting in San Francisco. Dr. Pasinetti's early findings suggest that there is a specific protein found in the spinal fluid that seems to be unique to patients with ALS. The amount seems to be related to the severity of the disease. The identity of the protein is not known at present but efforts to characterize this protein are well underway. Dr. Pasinetti's research is sponsored in part by the Muscular Dystrophy Association. We are pursuing additional studies of this protein and other potential biomarkers in our clinical studies at the MDA/ALS Program.

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The Beginning of the ALS Clinic

By Eileen Kellner

A long time ago, in 1976, a doctor named James Carascio came into my office in the Annenberg Building to tell me that his department chairman, Dr. Yahr had asked him to set up a clinic program for ALS patients. Together, he and I identified a space for it and I was able to recruit Sarah Caliman, who recently retired from Mount Sinai, to help him run the program. The program grew quickly and it became apparent almost immediately that it needed to be broadened. Patients who were unable to find help elsewhere in the country were calling for appointments. In a very short time, we received help from several other departments at Mount Sinai, including Social Services, Nursing, Home Care, Psychiatry and Rehabilitation Medicine.

There was much innovation. Occupational Therapy had feeding aids and other assistive devices stocked in their clinics ready to give to patients at the time of their visits. Family support groups were formed. Everyone, including patients and their families, worked together. It wasn't long before some of the family members came forward to Dr. Carascio and asked him to write a funding proposal to help the program and add a research component. Shortly after, the research program started.

I've had the pleasure of working with Dr. Dale J. Lange even before he came to Mount Sinai and I have met with some members of the interdisciplinary team. The MDA/ALS Team has done a lot to add to the service that began long ago and now, this newsletter for patients and families. It is important that the collaborative partnership among health care professionals, patients and families continues to fight this disease.

Research Update in Brief

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The MDA/ALS program at Mount Sinai School of Medicine is also conducting therapeutic trials in patients with multifocal motor neuropathy, a disorder that may look like a form of ALS (lower motor neuron form) but in fact is a peripheral neuropathy. We are trying to find better ways to identify patients with this disease.

A new clinical trial using an FDA approved medication that is used to treat other ailments, Disulfiram (Antabuse), is also being conducted at the MDA/ALS Program. We are currently involved in a safety trial and a larger double blind trial is planned after that study is complete.

On the last page of this publication is a listing of the clinical trials for which we are currently recruiting subjects and the contact information if you are interested in joining.

From our OT and PT

This column is written as a resource on adaptive devices and techniques to compensate for muscle weakness and help perform activities of daily living for people with neuromuscular diseases and their families.

Helpful Hints and Adaptive Devices

It is easier to hold a larger object if your hands are weak. Similarly, if your legs are weak, it is easier to stand up starting from a high surface than from a low one.

Good Grips Utensils – These cost roughly \$5 to \$8. They are light-weight and can easily be placed in the dishwasher to be cleaned.

Button Hook – This is a small device designed to perform the fine movements of buttoning one's shirt by weaving it through the hole to grasp the button. Cost is \$8.

Raised Toilet Seat – This item provides an increase of 5 to 6 inches of the overall seat height of the toilet, making it easier for the individual with weakness in the legs to stand from a seated position. The price of this item depends on the number of parts. A raised seat with handles and a clamp runs about \$75, whereas one that is simply placed on the toilet costs approximately \$20.

Transfer Tub Bench – The benefit of a transfer tub bench over a bath seat is that it eliminates the need to step into the tub. The individual simply sits on the bench, then swings their legs into the tub. This is beneficial if one has weakness in the legs or impaired balance. The price of this item depends on the materials used on the support surface and the feet. A padded bench with suction cups at the feet runs about \$175. A standard non-padded transfer tub bench costs \$120.

Grab Bars – These are usually mounted to the wall in a horizontal, vertical, or diagonal configuration. The price of these depends on the size of the bar, the materials used, and the texture of the surface. If you do not own your residence, it is important to speak to your landlord prior to installation of these items. When installing them in your own home, make sure they are being secured into the support beams of the house.

The above items can be ordered through a catalog, online, or at your local pharmacy. Under certain circumstances, an evaluation by an occupational therapist is recommended and a prescription from a doctor is required. For further information or examples of devices, you can go online to www.sammonspreston.com. Questions regarding equipment specifications can be directed to Jenny Lieberman, MSOTR/L at 212-241-7404.

2004 ALS Leadership Development and Clinical Conference

The 2004 ALS Leadership Development and Clinical Conference sponsored by the ALS Association (ALSA) was held March 12-13, 2004 in Woodland Hills, CA. The conference was well attended by healthcare professionals, patients and their families, researchers, and volunteer staff.

The clinical portion of the conference included two different workshops, one on a multidisciplinary approach to pulmonary care and one on strategies to improve quality of life, as well as networking and roundtable discussions by professional disciplines on a variety of topics.

The keynote speaker was Terry L. Wise, J.D. presenting "Rarely Heard Insights and Choices for Hope: A Caregiver's Perspective." Ms Wise was widowed at 35 following the death of her spouse from ALS. She discussed new perspectives on the impact of ALS, techniques that help break destructive patterns between patients and caregivers and skills to meet the needs of those affected by ALS and long-term care giving.

The quality-of-life track included presentations on three important dimensions that can all improve the emotional well-being of persons with ALS and their families: social support networks, the expressive arts, and spirituality.

Social Supports

Lance Christian, MSW, Social Worker from the ALSA Oregon and Southwest Washington chapter led the presentation and discussion on how important it is for ALS patients and caregivers to access social supports whether it be family and friends ("intimate supports"), professional resources ("formal supports") or religious sites, clubs, organizations, support groups ("community supports").

Mr. Christian stressed that the most vital step to helping a patient or caregiver achieve this is to make an assessment of their existing social environment, identifying their needs/wishes and looking for areas of strength and deficit. The next step is to use what he called an “ecological approach,” looking at where the different social supports overlap and helping patients and caregivers determine ways to improve their areas of deficit and maximize the areas of overlap. By doing this, patients and caregivers can be more interactive with others who they feel are important in their lives, have a more positive mood and feel less isolated.

The St. Louis area chapter presented a model of offering additional avenues of support to patients and caregivers living in rural areas who, due to geographical distances, are unable to attend support groups regularly. For families dealing with ALS for five years or more, a family mentoring program was developed in which families are matched based on demographic similarities and maintain telephone contact as often as they wish. At the end of the calendar year, ALSA hosts a banquet where families can meet each other in person and listen to various educational presentations. The participation in this program has grown tremendously over the years and has benefited many people by allowing them to share and learn from others facing similar challenges.

Expressive Arts

Denie Whalen, OTR, Expressive Arts Therapist in the ALS Center in Albany, New York, shared some of her personal experiences working with persons with ALS, illustrating the ways in which the arts can respond to the circumstances of our lives. She explained how precise, imaginative and heartfelt expression of feelings and emotions within the safe container of expressive arts can open the door to our deepest resources and broaden understanding of the possibilities inherent in the de-centering experience of being diagnosed with a serious illness.

Ms Whalen believes that the arts are an undervalued resource and that once people are given the tools and a nurturing environment, they allow themselves to grow. It is a shaping process that allows people to use their imagination and get in touch with their inner voice. The ALS Center in Albany successfully uses the expressive arts both in individual and group settings to help persons with ALS increase their artistic skills and intuition. The program offers the opportunity to relax as well as reflect.

Spirituality

Susan Walsh, RN, from the Hershey Medical Center ALS Clinic, presented one of the components of the workshop titled “Intervention to Enhance Spirituality.” Ms Walsh reviewed the multiple research projects on spirituality and ALS that her clinic has conducted since 1998, inspired by the abundant research in other diseases suggesting a positive relationship between health and religious participation.

The results of a 1998 study involving 96 ALS patients showed that quality of life correlates *not* with strength or function, but with existential, psychological and support factors from the McGill Quality of Life Measurement. It also showed that quality of life correlates with the total score from the Idler Index of Religiosity. In 1999, a year-long study of 60 ALS patients yielded similar results, showing that overall quality of life remained high, even as physical function declined when the spiritual component was strong (in terms both private, i.e., as perceived by the individual, and public, as measured by participation in religious services). The study also showed that the existential or meaning factor plays a significant role in quality of life and that religious, existential and spiritual measures need to be included in ALS quality-of-life measurements.

A third study conducted with 49 ALS patients with data collected from five consecutive visits over a 12-month period concluded that the importance of spirituality increases as ALS progresses. Although there was no significance at point one, at three months there was a small but significant relationship, at six and nine months, the spirituality factor accounts for 15% of overall quality of life and at 12 months, for 25%. This study concluded that private (vs. public) spirituality religiosity is the dominant factor at 12 months.

In 2003, the Hershey ALS Clinic validated the religious domain by conducting a multi-site study involving seven ALS clinics. It showed that religious factors (perceived religiousness, prayer, religion as a source of comfort and practices in the home) play a significant role in quality of life. Currently the Hershey ALS Clinic is conducting a pastoral care intervention study where they are assessing interventions developed based on their research, offering pastoral care in clinic, prayer cards, follow-up calls and referral and support to local church community.

ALS: Moving from a Fatal to a Chronic Disease

The closing plenary session, entitled “Conference Wrap-up and Future Directions: ALS as a Chronic Disease,” was conducted by Mary Lyon, RN, MN, Vice President, Patient Services, ALSA National Office. After presenting highlights of the two-day conference, the focus of the session was on changing the frame of reference of ALS from a fatal disease to a chronic disease. This changing view is due to the promotion of symptom management, data on use of NIV, PEG and LTMV, increasing scientific understanding, the use of biomarkers, treatment development, and early diagnosis and treatment initiation.

The session discussed a model for managing chronic diseases and the lessons that can be learned from multiple sclerosis, which is now considered a chronic disease. There was an identification of the challenges that clinicians and others helping people with ALS will face in the future as ALS becomes more of a chronic disease, as well as recommendations as to how to plan for and prepare to meet these challenges.

On Caregiving

This column is written by Meryl Kauffman Houghton, primary caregiver for her husband, Dr. Alan Houghton who was diagnosed with ALS 11 years ago. Meryl regularly attends the MDA/ALS Educational/Support Group at Mount Sinai, conducts thorough research on resources for her husband and has extensive care giving experience.

Question: What is it like to grow up with a parent with ALS?

Answer: Ten years ago I watched my husband Alan walk across our living room. He had recently been diagnosed with ALS, and the disease was steadily draining the strength in his arms and legs. I was at the other side of the house helping our 9-year-old son. Alan didn't look stable, and he was teetering from side to side. I couldn't reach him quickly so in a panic I called out to our 6-year-old to keep an eye on Dad. Alan tripped and fell as my son stood there with his mouth wide open. We learned two important lessons that day. Loafers and a foot drop are not compatible, and only assign tasks to children that they are capable of handling.

As children grow, their needs and perceptions of illness change. When our two boys were young, up to age 11, they couldn't wait for weekends to prepare a breakfast of toast with butter and honey for Alan. Putting shoes and socks on Dad was not a favorite, but they would argue over who would get the chance to drive the scooter or wheelchair out of the bedroom. On weekends, the boys would go with Alan in his wheelchair to the park where Alan would kick a soccer ball with the footpads of the wheelchair, and watch the boys climb on rocks and ride on the carousel. Afterwards they would share sodas and pretzels. Our 9-year-old was entrusted with the cell phone to call me if there was an emergency. Luckily we never had a problem.

When the teenage years arrived, the boys were not as eager to help. They were busy developing their own identities and tentatively exploring independence from their parents. Because they were often off with their friends or in their rooms behind closed doors, they were not readily accessible. Still, they were expected to be involved with Alan's care. Some of their tasks included restarting a frozen computer, loading software onto a computer, setting up books on a reading stand, performing range of motion exercises and scratching itches. We sometimes used portable monitors to allow Alan to communicate with us in other parts of our home. This was particularly useful on Saturday mornings when I went shopping. A monitor would be placed in one of the teenage boy's rooms while they were sleeping late. If Alan's computer crashed then our son could help.

Last summer, Alan spent almost three weeks in the hospital. Although the nurses and aids were always helpful, they were also very busy. In this situation Alan was very vulnerable. Because he was unable to move, he could not push a call button for assistance. Each of us took turns spending hours by his side. Nights were particularly difficult. Both boys were very concerned and spent long periods of time with Alan, making sure he was comfortable. Our 19-year-old son spent a complete night with Alan, sleeping on an uncomfortable chair, so that Alan would not be alone. Our sons' support during this difficult time was very important and inspiring. Alan recovered completely and the boys slipped back into their old routines, trying to avoid scratching Alan's itches.

Our sons have played an important role in the care of Alan although this role has changed as they have grown. On weekends they have been able to cover Alan for a few hours so that I can do errands, go shopping, or attend a ballet. However, we had to give them the appropriate space and freedom as they grew older and began to take on the challenges and responsibilities of growing into an adult. At each stage of their development, their participation in Alan's care brought us all closer together.

We look forward to answering your questions on care giving. Please submit questions to The MDA/ALS Beacon editor at nancy.xenakis@mssm.edu or Box 1137 Mount Sinai Medical Center, One Gustave L. Levy Place, New York, NY 10029.

MDA In May

May was ALS Awareness Month and the Muscular Dystrophy Association (MDA) is spreading the word about the disease and what MDA can do to help. Since 1950 MDA has allocated more than \$155 million in ALS research and medical care and has made great progress in the search for causes and treatments for ALS. MDA maintains 31 MDA/ALS clinics, one of which is our very own ALS Program here at Mount Sinai.

At MDA's Manhattan District office, we kicked off the month of May with the Empire State Building being lit up in MDA Blue on May 6. A press conference accompanied the lighting to get the word out about ALS Awareness Month and MDA's dedication to the fight against ALS.

To continue the ALS Awareness Month Activities, our main event was "The ALS Symposium and Vendor Expo," held on May 16 at The Clark Conference Center located in The Milstein Building of Columbia Presbyterian Hospital. Featured speakers included experts in the area of ALS research, respiratory care, augmentative communication and caregiving issues. In addition, vendors from leading health-care and equipment companies were on hand to demonstrate the latest in assistive equipment and services.

MDA Manhattan also offers an opportunity to get involved in raising ALS awareness. Through our ALS Awareness Cards program, you can pass along the word about the disease and MDA's involvement in the fight.

In the past three years MDA's Wings over Wall Street® has raised over \$3 million for ALS research. This year's event will be held on September 29 at The New York Marriott Marquis in Times Square. There are numerous opportunities to get involved with our mission to find a cure.

For more information on any of the above items as well as any of MDA's services please contact Leah Bailin, MDA/ALS Health Care Service Coordinator (212) 689-9040 or email newyorkcityalservices@mdausa.org.

What's on Your Mind?

Q: I understand that recently there has been a change in social security disability payments for people with ALS. Can you tell me more about this?

A: The Social Security Administration recently approved the inclusion of presumptive disability for people with ALS. This ruling is effective immediately and means that people with ALS will no longer have to prove that they meet the Social Security definition of "disabled." In place of the previous proof required (a physician's certification that there was significant bulbar dysfunction or diminished use of two limbs) the Social Security Administration now states that a diagnosis of ALS is sufficient to be considered "disabled."

Unfortunately, this ruling does not mean that everyone diagnosed with ALS will be eligible for Social Security Disability Income (SSDI). In addition to being disabled, the applicant must still have a sufficient work history of paying Social Security taxes in order to be eligible for SSDI. The ruling also does not mean that one can get SSDI immediately upon being diagnosed. People with ALS still need to wait the five -month period after applying before they will begin receiving their benefits.

Due to this recent ruling, people who are diagnosed with ALS and wish to stop working, either because they feel that it adversely affects their health or because they wish to spend more time focusing on other aspects of their lives, can now apply for benefits at the time of their diagnosis.

Although the Social Security Administration makes an effort to communicate all updates to its regulations to its workers, it is possible that local Social Security offices may not be aware of this ruling. If you are in the process of applying for SSDI, let your local office know that the new regulation is included as amendments to parts 404 and 416 under Title 20 of the Code of Federal Regulations and was effective as of August 28, 2003. This will help them access the information and process your request more efficiently.

To apply for SSDI or to find out any information regarding Social Security benefits, call 1-800-772-1213 and select the option to speak to a representative. This Social Security representative can inform you of your eligibility status for SSDI based on the length of your work history and if you qualify, can make an appointment with your local Social Security office to process an application.

We look forward to hearing what's on your mind. Please submit questions to the MDA/ALS Beacon editor at nancy.xenakis@mssm.edu or Box 1137 Mount Sinai Medical Center, One Gustave L. Levy Place NY NY 10029

What's Happening in the MDA/ALS Educational/Support Group?

The Educational/Support Group at Mount Sinai Medical Center, which began in May 2003, is a forum where patients and caregivers meet on the first Wednesday of every month from 6-7:30pm to learn about a different educational topic and then share their experiences, thoughts and feelings as a group. Dale J. Lange, MD, Director of the Program is also available at the beginning of each group for a question and answer session. A light dinner and a table of educational materials are provided. Some educational topics that have been presented to date include:

- Questions and Answers with MDA/ALS Program Medical Director
- Occupational Therapy: Assistive Devices for Activities of Daily Living
- Family Dynamics and Issues Confronting Caregivers
- Therapeutic Trials: Why, How to, and Where we are
- Safe and Comfortable Travel for the Physically Challenged
- The Cost of Care: Caregivers' Financial Concerns
- Mobility and Transfer Training
- Planning for your Future: Financial and Medical Issues
- Review of the Proceedings from MND and ALS Conferences
- Maintaining Nutrition with ALS and MND
- Respiratory Support Issues and Related Equipment
- Accessibility in and Around the Home

Please call us at 212-241-6049 if you are interested in receiving future group mailings or if you have an idea for an educational topic. You can also find out about our groups by visiting our message board at: www.mssm.edu/neurology/neuromuscular/als

Visit Us Electronically

www.mssm.edu/neurology/neuromuscular/als and link to our new message board.

It contains three forums:

- Educational/Support Group
- Questions Regarding our Services
- General Discussion

Clinical Research Studies at The Mount Sinai Medical Center

The following are clinical trials in which we are currently enrolling subjects. For more information please contact 212.241.8984.

- IVIG (Intravenous Immunoglobulin) in Lower Motor Neuron Syndromes (Motor Neuropathy)
- Multicenter Study Using the "VEST" (High Chest Wall Frequency Oscillation) in ALS
- The Safety and Tolerability of Disulfiram Treatment in ALS

Helpful Reference Materials

Journals/Newsletters/Websites:

- Quest
- MDA/ALS Newsletter
- www.mdaua.org
- www.mssm.edu/neurology/neuromuscular/als
- www.mountsinai.org/msh/clinical_services/wecope
- Bob Broedel's online ALS Digest: to register: bro@met.fsu.edu, to review back issues: www.alslinks.com

Books:

- "Waking Up" by Terry L. Wise, JD
- "On Any Given Day" by Joe Martin
- "Anatomy of Hope" by Jerome Groopman, MD

From the MDA:

- ALS: Maintaining Mobility
- ALS: Meals
- ALS: A Caregivers' Guide

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