

THE UNIVERSITY OF **Miami** ALS Center NEWSLETTER

KESSENICH FAMILY MDA

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Kessenich Corner



Ramon Antelo, Rep. Ileana Ros-Lehtinen & Dr. Walter G. Bradley

2004 National ALS Advocacy Day

The ALS Association Florida Chapter and President, Dara

Alexander, organized a

delegation from Florida to advocate for ALS in congress on Monday the 17th of May, which was ALS Awareness Day this year. The delegation was the largest ever at about 25 people including patients and relatives. I enjoyed making the rounds of our Congressmen and Senators with Ramon Antelo, whose father, sadly, has ALS. We were able to meet with Representatives Lincoln Diaz-Balart, Ileana Ros-Lehtinen, Robert Wexler and Clay Shaw, and met with Staff Assistants of Senators Bob Graham and Bill Nelson. Delegates from the other parts of Florida were able to meet with their congressional representatives. We emphasized the importance of increasing the appropriation to the National Institutes of Health by 8% in this year's budget, which will only just keep pace with the rate of inflation in medical research costs. We also advocated for continu-

ing the research funding for the Department of Defense, part of which is going to research into ALS because of the increased risk in Gulf War veterans. We also advocated for co-sponsorship for the Rachel Grainger Act of 2003 which calls upon the Social Security Administration to expedite the processing of claims of terminally ill patients so that they will live to see the benefits that they desperately need. We received a courteous hearing at all of our visits, and I believe that they will result in increased support for ALS patients and research.



Dr. Walter G. Bradley & Ramon Antelo

It is clear that ALS Advocacy Day and the presence of so many patients and loved ones attending

every year is having an important effect on expanding awareness of ALS in Congress. I urge all of you to think of coming to Washington next year.

Walter G. Bradley, DM, FRCP
Professor and Chairman
Department of Neurology



A new gene causes a rare form of ALS

Ashok Verma, MD, DM

Approximately 10% of ALS cases are familial (FALS). A

genetic fault is the underlying cause of impaired function and premature death of motor neurons in FALS. Motor neurons are located in the brain and spinal cord and their loss causes ALS. Although rare, careful study of these FALS cases may provide clues to the cause(s) of more common nonfamilial or sporadic forms of ALS.

A team of researchers led by Dr. Chance in Seattle recently reported that a defect in a newly discovered gene SETX causes a juvenile-onset slow form of ALS. Two other genes have also been linked to FALS in previous studies: SOD1 and Alsin. Approximately 20% of FALS cases are associated with SOD1 gene defects. We all have two copies of each of these genes in our genetic blue print. Only one of the two copies of SOD1 or SETX when flawed can cause ALS. In case of alsin, two abnormal copies are required for ALS. How defects in these genes cause death of motor neurons and ALS is the area of intense current research.

SETX genes help cells to manufacture a protein "senataxin" which belongs to a family of molecules known as "helicases." Helicases normally help the cells to interpret and process genetic instructions along the DNA-RNA pathways.

Smooth flow of instructions from DNA to RNA to protein manufacturing is needed for cell's health. When senataxin protein is flawed, as it is in some juvenile ALS families, this smooth genetic instruction is disrupted, resulting in gradual death of motor neurons.

Interestingly, defects in both copies of senataxin have been reported to cause another progressive neurological disease known as AOA2 (ataxia-ocular apraxia type 2). Many cases of AOA2 also show degeneration of motor neurons.

Although the more common form of nonfamilial ALS differs from senataxin-associated ALS, as it is not clearly genetic, it begins in late middle age, and it progresses rapidly to paralysis and death. Both diseases cause selective death of motor neurons. The senataxin discovery is important for two reasons. First, the linkage of a helicase gene to motor neuron death puts another important piece in the puzzle of ALS. It will likely help understand the mechanism or steps of cell death in ALS. Second, it may provide a potential target for the development of treatment for this devastating disease.



A Day of Pampering for a Teen with ALS

The Make-A-Wish Foundation made January 10, 2004, a day that 17 year old Elizabeth Capehart would never forget. On October 16, 2003, Elizabeth was diagnosed with ALS (Amyotrophic Lateral Sclerosis, or "Lou Gehrig's Disease") at the age of 17. This vibrant and active teen has been forced to deal with a terribly debilitating disease, but for a day was able to forget about it and was treated like a princess.

Elizabeth has the same concerns as any other teen- being a 17 year old is hard. Due to mounting medical bills and an uncertainty about the future, her family has not been able to spend money on the 'extras' that are essential to teen living. Elizabeth requested a shopping spree in the local area mall from the Make-A-Wish Foundation. The Make-A-Wish Foundation pulled together a terrific day for Elizabeth, her mother, two sisters and



best friend.

The Foundation sent a white stretch limousine to chauffeur the party out for the special day. The limo took her and the gang on a ride through town on the way to the local mall. She was greeted at the front door by the staff of Burdines armed with greeting signs and balloons.

First, Elizabeth was escorted to the boutique for a personalized makeover, complete with a staff dedicated to perfecting her hair and make-up. After leaving the boutique, Elizabeth was escorted downstairs to "Shop Till She Dropped." She had the option to choose from perfumes, handbags, clothes, shoes, home accessories and much more. She



was then escorted around the mall to any store she desired. This included stops at Foot Locker, Jewelry Stores, Victoria's Secret and Gadzooks to name a few. Later a mall associate greeted her with a mall gift certificate for her to enjoy another day.

To keep up her strength and spirits, the limo chauffeured the crew to Red Lobster for a lunch break. The group was led to a table set and decorated for the party. Elizabeth and the crew were treated to a fabulous lunch.



After lunch, the limo drove the group back to the mall for another round of shopping. There, Elizabeth was presented with personalized gifts from "Things Remembered." She finished up her day purchasing a few more items and was then taken home, making the mandatory detour to the local hang out to show off for the crowd.

Elizabeth's day was complete with several photo-ops and reels of video to remember her special day. She was extremely pleased with her day of pampering, especially because she was able to enjoy it with all of her family and friends. Elizabeth is very grateful to the caring people from the Make-A-Wish Foundation that worked to make her wish come true.



**Her loving sister,
Wendy L. Joseph
Dixie.Joseph@nnsa.doe.gov**

HURRICANE SEASON

Hurricane season is from June 1 to November 30. Here's some information on what to do if a hurricane is heading your way and a disaster plan.

Before a hurricane comes knocking:

- Check your insurance. If you live in an area that could be affected by a hurricane's storm surge, make sure you have flood insurance.
- Prepare to take with you a box with your medical insurance papers, homeowner's insurance papers and other valuable papers. Keep a list of prescribed medicines and photocopies of prescriptions, etc. Place valuables and personal papers in waterproof containers and store them in a safe place.
- Plan a meeting place. Make sure that everyone in your family knows where to meet, a relative's house, for example, - to prevent family members from becoming separated.
- Pick an emergency contact.
- Develop an emergency communication plan in case family members are separated. Ask an out of state relative or friend to serve as the family contact. Make sure everyone in the family knows how to reach this contact person.
- Be ready to evacuate. When an order comes to evacuate your area, be prepared. Have your car fueled up and your disaster kit ready to go.
- If you need assistance evacuating to a Special Needs Evacuation Center, you will need to register in advance. Please contact your County Emergency Management Office or log on to their website: www.co.miami-dade.fl.us/oem

Miami-Dade	305-468-5400
Broward	954-831-3900
Monroe	305-296-4033
Palm Beach	561-712-6400

- Emergency Evacuation Assistance Program (EEAP): Many of your neighbors may need assistance with their daily living, and a disaster occurring makes them even more vulnerable. OEM maintains a registry of Miami-Dade residents that will need assistance evacuating before or after a disaster. IT IS NOT JUST FOR HURRICANES! Any sudden incident that may require evacuations could trigger the need to help some of these residents with special needs.

- Special Needs Evacuation Centers (SNECS): These are opened as places of safety for residents on the registry. When eligible registrants are provided with transportation, additional medical personnel at the SNEC and placement at safe facilities until the threat from the disaster is over.

- Anyone who needs assistance, including help with activities of daily living and electrically dependent medical equipment, should register for the EEAP as soon as possible. Contact them via website: www.miamidade.gov/oem or call directly to receive an application:

Miami-Dade	(305) 305-513-7700 305-468-5402 (TDD) 954-537-2888
Broward	954-537-2882 (TTY) 305-292-4591
Key West	305-292-4418 (TTY) 305-743-0079 ext. 4591
Middle Keys	305-743-0079 ext. 4591
Upper Keys	305-852-1469 ext. 4591
Palm Beach	561-712-6400

During a hurricane watch:

- Stay aware. Listen to radio or television hurricane reports. Persons assigned to Special Needs Evacuation Centers should listen to the media and determine if they must evacuate and what time the centers will open. Remember, do not go to the shelter until you hear an announcement that it is open.



Rumor Control Lines (IN DISASTER SITUATIONS ONLY):

Hurricane Hot Line 305-229-4483
www.nhc.noaa.gov

Answer Center:

Miami- Dade 305-468-5400

Citizen Information Line:

Broward County 954-831-4000

Emergency Information Center:

Palm Beach 561-712-6400

Emergency Information:

Monroe 800-955-5504

State of Florida Emergency

Information Line 800-342-3557

TTY: 800-226- 4329

- If you live in a mobile home or are **ELECTRICALLY DEPENDENT**, you should evacuate to the closest hospital for any category of tropical storm or hurricane. Do not stay there under any circumstances. Mobile homes are unsafe in hurricane conditions, no matter how well fastened to the ground.

- Check your emergency supplies, turn your refrigerator and freezer to the coldest setting and open them only if necessary. Store drinking water in clean bathtubs and bottles. Figure one gallon of drinking water per person, per day.

- Prepare your vehicle. Fill your fuel tank early. Gasoline may not be available for days after the hurricane due to power outages. Make sure to have adequate cash on hand.

- You should lock and board up windows. Stow away small objects that could cause damage in high winds. Remove outside antennas. If you are going to ride the storm out elsewhere, turn the utilities off.

During a hurricane warning:

There are two types of official warnings:

1) A hurricane watch: a hurricane may strike within 24-36 hours.

2) A Hurricane Warning: a hurricane is expected to strike in 24 hours or less.

- Listen to your radio or television for official instructions

- Stay inside the house away from windows and

glass doors. Secure windows and shutters

- If you've been ordered to evacuate, leave as soon as possible. Make sure you tell someone where you are going.

During a hurricane:

- Stay away from windows and doors even if they are covered.

- Take a battery-powered radio, TV and a flashlight with you to your place of refuge.

- Close all interior doors.

- Go to an interior room such as a bathroom, closet or under the stairs.

- Lie on the floor under tables or other sturdy objects.

After a hurricane:

- Remain indoors until the official "all clear" is given.

- If you have evacuated, do not return home until officials announce your area is ready.

- Boil water immediately after the hurricane passes. Water supplies may be contaminated during a hurricane.

- Use only your pre-stored, dry or canned food.

- Do not touch fallen or low-hanging wires of any kind and under any circumstances.

- Treat every power line you see as deadly.



Sweet Pepper Puree

Ingredients

- 5 medium-sized red, yellow or green bell peppers, seeded and cut into one inch chunks
- 6 cups cold chicken broth
- 1 medium-sized red onion, cut into one inch chunks
- 1 medium boiling potato, peeled and cut into one inch chunks
- 8 bay leaves
- 1 teaspoon dried thyme, crumbled
- 6 scant teaspoons olive oil, salt and ground pepper

Procedure

1. Soak pepper in cold water 30 minutes.
2. Drain peppers and combine with broth, onion and potato in heavy medium saucepan over medium heat.
3. Cover and simmer until vegetables are tender, about 45 minutes.
4. Puree soup through fine disc of food mill or food processor. Return to saucepan.
5. Wrap 2 bay leaves, thyme and clove in cheesecloth and add to soup.
6. Season with salt and pepper.
7. Bring to simmer and cook until thick, stirring occasionally, about 25 minutes.
8. Soak remaining bay leaves in luke-warm water to cover 5 minutes and drain.
9. Discard cheesecloth package.
10. Ladle soup into bowls.
11. Top each with bay leaf and drizzle with olive oil in "C" pattern.
12. Serve hot or cold.



Orange Pudding Cake

Ingredients

- 1 cup honey
- 2 tablespoons butter
- 1 1/2 tablespoons orange rind, grated
- 4 eggs, separated
- 1 cup flour
- 1 cup milk
- 1 cup orange juice
- 8 packets of Equal

Procedure

1. Mix honey, butter and orange rind together.
2. Add egg yolks one at a time, beating well after each addition.
3. Add flour, milk and orange juice gradually.
4. Beat egg whites in separate bowl, add 8 packets of Equal and beat until stiff.
5. Fold into orange mixture.
6. Place mixture in buttered casserole and bake at 350 degrees for 1 hour.

Announcement

THE KESSENICH FAMILY MDA ALS CENTER LOAN CLOSET IS OVERSTOCKED WITH MEDICAL EQUIPMENT FOR ANYONE THAT NEEDS THEM. WE HAVE ELECTRIC WHEELCHAIRS, CANES, WALKERS, SHOWER CHAIRS, AND MUCH MORE. IF YOU HAVE ANY QUESTIONS OR FOR A LOAN APPLICATION, PLEASE CALL **(305) 243-7400** AND SPEAK WITH GINNA OR GRACIELA.



Important Numbers

- Kessenich Family MDA ALS Center 305-243-7400
 1-800-690-ALS-1
 www.miami-als.org
- Muscular Dystrophy Association (National Patient Information) 1-800-572-1717
 www.mdausa.org
- St. Petersburg 717-576-5202 or 1-800-393-8552
- Palm Beach Gardens 561-242-5084 or 1-800-289-0535
- Miami 305-717-9937 or 1-800-572-0085
- Broward (954) 757-4357 or 877-970-9696
- ALS Association 1-800-782-4747
- ALS Association in Florida 1-888-257-1717
 www.alsa.org
- National Caregiving Fdn 1-800-930-1357
- National Family Caregivers Assn 1-800-896-3650
 www.nfcacares.org
- Foundation for Hospice and Homecare 202-547-6586
- National Hospice Org. 1-800-658-8898
- A.D.E.L.A. Asociación Española de Esclerosis Lateral Amiotrófica
 www.advernet.es/atlela/index.htm
- Social Security Online 1-800-772-1213
 www.ssa.gov
- The Feeding Gastrostomy Information: www.iinet.net.au/~scarlfam/gtube.html
- ALS Digest (Bob Broedel): To subscribe, please e-mail to bob@huey.nict.fsu.edu

If you need a referral to one of our satellite centers, please call 305-243-7400 or 1-800-690-ALS1

NOTES:

* We are seeking volunteers to assist in running the ALS Center and to assist patients and families at home. Please call 305-243-7400. This could be a health care professional or even a family member who has had experience with ALS patients and now has time to volunteer.



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**The Kessenich Family
MDA ALS Center**

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Suite 701
Miami, Florida 33136

ALS Support Group Dates For The Year 2004

Calendar

- Saturday, July 10
- Saturday, August 14
- Saturday, September 11
- Saturday, October 16
- Saturday, November 13 (Thanksgiving Luncheon)
- Saturday, December 11 (ALS Symposium)



The ALS support groups will be held from 12:00 p.m. to 1:30 p.m. at the University of Miami Hospital and Clinics, 1475 NW 12th Avenue, Room #1301.

We hope to see you at the meetings where we will hear talks by our doctors and therapists, and different healthcare companies and providers will give demonstrations of their newest products and services.

*The above dates are subject to change and in the event of rescheduling, we will inform you via mail. Please call Ginna or Graciela at (305) 243-7400 if you have any questions, or you can visit us at www.miami-als.org.

ALS FOF Golf Tournament (Tampa) Oct. 23 for more info call 305-243-1005

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