SUMMER 2010

The AlphaNetter

The *AlphaNetter* is a publication of AlphaNet, Inc.

AlphaNet, Inc.
2937 SW 27th Ave., Suite 305
Coconut Grove, Florida 33133
800-577-2638, Fax: 305-442-1803
www.alphanet.org

Harmonica Instrumental in Breathing Tune-Up

Making music is making breathing easier for Alpha John Shook and the other members of the Harmonikatz, the core group of harmonica aficionados in the Florida Celebration Health pulmonary rehabilitation program.

"Initially, we were trying to do something to build up our diaphragms and intercostal muscles, which you need for proper breathing. Our diaphragms were kind of sluggish," John says. "Our instructor, Patricia Ross, recognized that some musical instruments can help us practice proper breathing. She outfitted our group with $4 harmonicas from Cracker Barrel and brought in an instructor to show us how to use them."

John laughs as he describes the cacophony first produced by a room full of amateur musicians. "It's good for our health and we laugh a lot along the way. We've learned to make loud mistakes and, like anything, we've improved with practice."

According to respiratory therapist, Jeff Fraser, the breathing techniques used to play the harmonica are very similar to some standard exercises used in pulmonary rehabilitation programs for COPD patients. For example, COPD patients tend to get air trapped in their overextended air sacs, causing shortness of breath. Purse lip breathing helps patients to keep their airways open, to exhale longer and to expel air from of their lungs with their abdominal muscles. A similar type of resistance is created when exhaling through pursed lips to produce notes on the harmonica.

Many COPD patients also develop flattened diaphragms, which do not constrict as efficiently and cause patients to use accessory muscles in the chest, neck, arms and shoulders to help them breathe. Harmonica play requires diaphragmatic breathing which strengthens the diaphragm by inhaling through a restricted space. During inhalation, the diaphragm contracts, creating pressure that allows more air to be drawn into the lungs.

"To teach our patients breath control, we have them focus on playing a single note and trying to fine tune it much like they would tune in a radio station," Patricia Ross, lead respiratory therapist, explains. "When patients utilize their diaphragms properly, they will hear a rich, strong sound. Although most of our patients have never played an instrument, they have become proficient in a variety of harmonica songs ranging from 'Mary Had a Little Lamb' to 'The Battle Hymn of the Republic'."

"One of the program's goals is to train patients in proper breathing technique and the harmonica has been an excellent learning tool," Jeff adds. "It helps patients to see improvement and provides an incentive to stick with the breathing pattern."

"Our breathing has improved and our mutual enjoyment has helped us to develop a strong support community," John says. "We are eager to share what we've learned with others."

Harmonikatz member John Shook plays his harmonica.

The Harmonikatz have performed for pulmonary rehabilitation programs, for COPD support groups, at parades and at other events in South Florida. Their goal in these performances is to entertain, encourage and educate other COPD patients.

*Pick up your **FREE HARMONICA** from the AlphaNet booth at the Alpha-1 Association National Education Conference.*

Understanding Your Genes

by Dr. Robert A. Sandhaus, MD PhD, FCCP,
Medical Director, AlphaNet

The following questions were submited by a member of the Alpha-1 community.

If an individual has a genotype of ZZ, is the alpha-1 antitrypsin that escapes the liver able to protect the lungs at all since the protein is mis-folded?

The Z alpha-1 antitrypsin protein functions very well as an inhibitor of the neutrophil elastase that is thought to cause the lung disease of Alpha-1. Out of the 394 amino acids that make up the alpha-1 antitrypsin molecule, there is only one amino acid that is different in the Z protein compared with the M protein (normal alpha-1 antitrypsin). That one amino acid difference does allow the molecules of Z protein to stick together inside the liver cells, but the Z protein that makes it into the circulation seems to work quite well. That's why some people with ZZ Alpha-1 never get any lung disease at all.

Since the body makes small amounts of alpha-1 antitrypsin in places other than the liver, why not target those places for making more of the protein when trying gene therapy?

The vast majority of the alpha-1 antitrypsin in the circulation and the tissues comes from the liver. That's why, when an individual with Alpha-1 gets a liver transplant, their blood will have only normal alpha-1 antitrypsin in their circulation. There is good evidence that the cells that have been shown to make alpha-1 antitrypsin get clogged with abnormal protein just like the cells in the liver. Having said all this, it is interesting to note that the current gene therapy studies going on in Alpha-1 are aiming to turn muscle cells into alpha-1 antitrypsin secreting cells.

Alpha Support Group Visits Kotton Lab

In May, Alpha Joanne Mellady and other members of the Massachusetts Alpha-1 support group traveled to the Kotton Lab at the Boston University School of Medicine to learn about the roles of gene therapy, gene transfer and skin-to-stem-cell biology in Alpha-1 research.

The visitors were treated to a presentation by Dr. Darrell N. Kotton, Associate Professor of Medicine and Pathology and Laboratory Medicine, and his staff. Dr. Kotton explained that, in a recent study, four samples of Alpha-1 skin cells, taken from skin biopsies, were successfully reprogrammed into a novel population of stem cells called induced pluripotent stem (iPS) cells. The attendees had the opportunity to view these samples under a microscope.

This study is the first step in a much larger undertaking. The next step is to isolate the point at which stem cells differentiate to form individual organ cells, such as liver cells. Theoretically, if researchers can identify what happens to the liver cells at this stage to indicate that there is Alpha-1 antitrypsin deficiency, they could begin to correct the problem at its source. Dr. Kotton and his staff are also looking to engineer methods to transplant cells with healthy Alpha-1 genes into the liver or lungs.

"It was incredible to learn about the advances they are making on a cellular level," Joanne says. "One girl was creating liver cells from stem cells and all the cells were producing. It was especially interesting for

Healy Tees Up For Alpha-1 Research

This August, seasoned and amateur golfers alike will shoot for the green at the 2nd Annual Paul Healy Golf tournament in Halifax, Massachusetts. Alpha Bob Healy started this "working man's" golf tournament in memory of his brother Paul Healy, an Alpha, lung transplant recipient and avid golfer. Paul passed away in 2002.

"Paul tried to play every weekend. He always wanted to get in another round of golf. I see this tournament as a way to honor him, raise awareness and support research. Last year we donated $5,500 to the Alpha-1 Foundation in his memory," Bob says.

Bob was diagnosed with Alpha-1 in 2000. He heard about Zemaira at an Education day in Lebanon, New Hampshire and contacted his doctor to be put on product. In October of 2004, he began infusing Zemaira.

Since his diagnosis, Bob has been quick to make a mark in the Alpha-1 community. In addition to his golf tournament, Bob chaired this year's Celtic Connection, has been a member of Team Alpha at the "Escape to the Cape" Bike Trek and has participated in several research studies.

"The way I see it you have two paths you can take: resign yourself to this disorder or go out and see how you can make yourself better," Bob says. "I am in the gym everyday and I play golf about five times a week. I make an effort to participate in research studies whenever I can, because I know that contributing my time can help us gain valuable information about our condition and can eventually lead to a cure. That's why I host this tournament. In my mind every fundraiser brings us another step closer to finding a cure. If I can do something to help someone else have a brighter future, and a better quality of life, that's just another goal I wish to accomplish."

This year's tournament will be held on August 9th at the Halifax Country Club in Halifax, Massachusetts. The cost is $100 per person. Interested golfers of any ability can contact Bob via email at bobhealy125@msn.com

me, because I have recently been involved in a research study."

Joanne was diagnosed with Alpha-1 in 1990 and received a double-lung transplant in 2007. "My transplant has been successful, but I recognize the value of participating in research that may produce a standard cure for future generations. It's amazing to see the progress they are making. This visit has been such an encouragement."

Dr. Kotton will be presenting his research at the Alpha-1 Association National Education Conference in Orlando, Florida. For more information on his research activities visit www.kottonlab.com

Participate in a Research Study or Clinical Trial

Those willing to participate in research studies and clinical trials related to Alpha-1 Antitrypsin Deficiency may want to consider signing up for the Alpha-1 Research Registry.

The Alpha-1 Research Registry is a confidential database of individuals with Alpha-1 Antitrypsin Deficiency (Alpha-1) or a carrier phenotype located at the Medical University of South Carolina in Charleston, SC. The goal of the Registry is to facilitate Alpha-1 research by providing investigators with a group of Alphas and carriers willing to consider participation in research.

For more information visit www.alphaoneregistry.org

Clinical Corner

Maintain Your Mobility

By Teresa Kitchen, BSN, RN, AlphaNet Clinical Nurse Manager

How much do you appreciate the flexibility that comes with driving your car, walking with a friend or spouse, preparing meals, traveling to visit family, and managing your personal care? All of these activities require a degree of personal mobility and increase your opportunities to interact with others and maintain independence. Personal mobility can play a significant role in improving your quality of life. And the choices you make now can influence your quality of life down the road. As the saying goes "if you don't use it, you lose it". Here are a few ideas to help you be proactive about maintaining and increasing your mobility.

- Make a list of the activities you enjoy and want to continue to be able to perform. Next to that list write what you need to do to maintain the ability to do them. If you don't know, consult your doctor.

- The best way to maintain or increase your ability to do an activity is to do it on a regular basis. Alphas need to be as physically active as possible, because the stronger your muscles are, the less oxygen you need to use.

- Identify tools that will help you monitor and increase your activity. The use of a pedometer (a device that measures your steps) or thera-bands (exercise bands) are both great ways to improve your strength and endurance and therefore increase mobility.

- Utilize medical equipment that can help you get around and perform daily activities. Wheelchairs, walkers and scooters can help Alphas maintain a more active lifestyle.

- Identify roadblocks that prevent you from participating in the activities you enjoy and create solutions by modifying the activity with support tools. Those using oxygen may consider a portable oxygen concentrator. There are many equipment options to choose from and it is best to speak with your home medical equipment company or oxygen supplier for their advice.

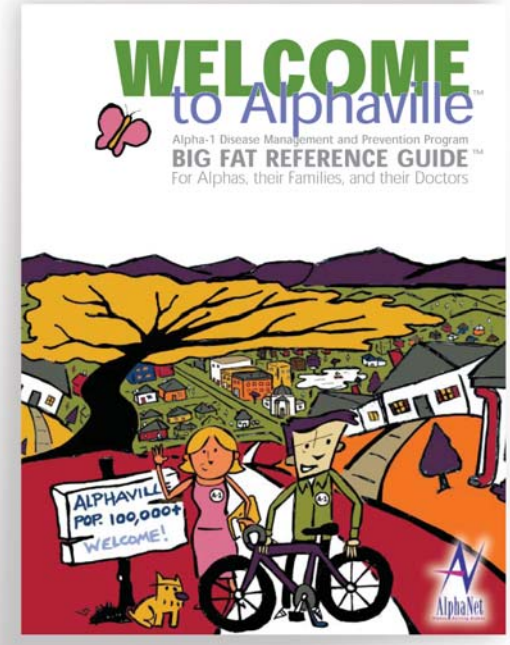
- Remember, for a medical support device to be covered in part or in whole by Medicare or private insurance, you should obtain a prescription from your doctor.

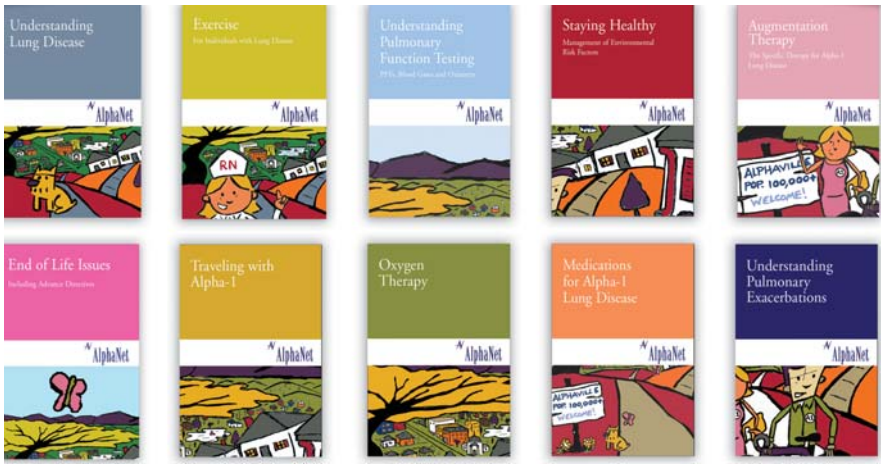
If you have a topic you think that all Alphas should learn about in the next issue of the AlphaNetter, please contact Lindsey Griffin at lgriffin@alphanet.org or 800-577-2638 ext.243

50 Screened at Alpha-1 Testing Day

In April, the American Lung Association, Pennsylvania State Hershey Medical Center and the Central Pennsylvania Alpha Support Group provided information about COPD and Alpha-1 and provided free testing for asthma, COPD and Alpha-1 Antitrypsin Deficiency at the Friendship Center in Harrisburg, Pennsylvania. Hershey Medical Center doctors, led by Dr. Timothy Craig, screened over 50 individuals. The screening consisted of spirometry testing and Alpha-1 testing using kits provided by CSL Behring and Talecris Biotherapeutics. CSL Behring and Talecris Biotherapeutics also provided beverages and breakfast snacks for the participants.

(Pictured right) Amy Czech of the American Lung Association and Jean McCathern, AlphaNet Coordinator (ME, VT, NH, CT, MD), work the information and check-in table at the Alpha-1 Testing Day in Harrisburg, Pennsylvania.





Stop by the AlphaNet booth at the National Education Conference for your free BFRG USB drive!

The Big Fat Reference Guide That Fits in Your Pocket

The Big Fat Reference Guide, the most comprehensive guide to living with Alpha-1, is now available in a new USB drive format. This up-to-date version of the BFRG is easy to transport and can be used on any computer with a USB port. The Skinny Little Reference Guide collection of 13 topic-specific guides is also included on the drive. Stop by the AlphaNet booth at the Alpha-1 Assocation National Education Conference in Orlando for your free copy. The BFRG and SLRG's can also be accessed online at www.alphanet.org.

AlphaNet is proud to support members of the Alpha-1 community with its unique focus on individualized needs, specialized health management programs and outcomes research.

For the tenth consecutive year, AlphaNet is pleased to provide scholarships for 40 participants to attend the Alpha-1 Association National Education Conference.

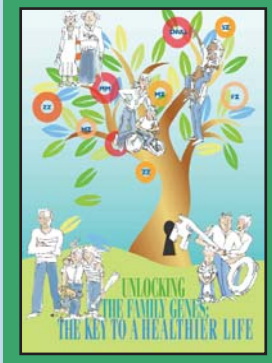
Community Calendars

Alpha-1 Association Education Days

co-sponsored by
the Alpha-1 Foundation

August 28
Des Moines, Iowa

September 25
Las Vegas, Nevada



Alpha-1 Association National Education Conference

June 11 - 13
Orlando, Florida

To find out more about conferences and support groups, visit the Alpha-1 Association website at www.alpha1.org or call toll-free: (800) 521-3025.

AlphaNet Employment Opportunity

Do You Have What It Takes?

Applications are currently being accepted for employment as AlphaNet Patient Service Coordinators. Interested Alphas should be dedicated, organized professionals who are able to travel and demonstrate strong computer and communication skills.

Send resumes to humanresources@alphanet.org

Alpha-1 Foundation Event Calendar

June 5 - Alpha Bike Run:
Mohawk Ramblers Motorcycle Club
Greenfield, Massachusetts

July 18 - Second Annual
Get the Scoop on Alpha-1
Johnston, Iowa

August 9 - Paul Healy Golf Tournament
Halifax Country Club
Halifax, Massachusetts

September 9 - Alpha-1 Golf Tournament
Greenwich, Connecticut

October 1 - 3 - Team Alpha-1
Escape to the Cape
Cape Cod, Massachusetts

November 20 - Alpha-1 5k Walk Miami
Miami, Florida

To find out more about these or other events in your area, log onto www.alphaone.org or call toll-free: (888) 825-7421, ext. 248.

AlphaNet
2937 SW 27th Ave, Suite 305
Coconut Grove, Florida 33133
800-577-2638 • www.alphanet.org
Return Service Requested