

GRADS Study Aims to Apply Genomics to Alpha-1

By: Robert A. Sandhaus, MD, PhD, AlphaNet Medical Director

The National Heart, Lung, and Blood Institute (NHLBI) of the National Institutes of Health (NIH) is funding a major research initiative called GRADS (Genomic Research in Alpha-1 Antitrypsin Deficiency and Sarcoidosis). The Alpha-1 component of the GRADS study is designed to look at the genes of the bacteria, viruses and fungi living in the lungs of individuals with Alpha-1 Antitrypsin Deficiency (Alpha-1) to help determine if these “germs” have genetic characteristics that affect the severity of the lung disease that many Alphas develop. While we know that environmental factors, such as cigarette smoke, play a major role in promoting lung disease in Alphas, we also know that Alphas with similar smoking histories can have wide differences in their lung problems. Some Alphas who have smoked have no lung disease, while others with minimal or no smoking history have severe emphysema. This study is designed to find out if the organisms living in the lung play a role in determining severity or if they could be markers that change, depending on disease severity.

GRADS investigators are based at eight study sites scattered around the U.S. and a study coordinating center at the University of Pittsburgh. The study sites include the Medical University of South Carolina, National Jewish Health in Colorado, Yale University in Connecticut, Johns Hopkins University in Maryland, University of Arizona, University of California - San Francisco, University of Pennsylvania, and Vanderbilt University in Tennessee (see contact information in right-hand column).

Participants in the study must have PiZZ Alpha-1 or be a PiMZ carrier. PiZZ patients can be receiving augmentation therapy or not. Those with PiMZ genotype cannot be receiving augmentation therapy. Participants' lung function must be good enough to tolerate a bronchoscopy – a procedure in which a small flexible tube with a light and miniature TV camera at the end is passed through the mouth down into the windpipe and then on into the bronchial tubes. The bronchoscopy is performed with a mild anaesthetic that puts the person into a light sleep and the procedure is done to wash the lungs with saline (salt water) to collect samples of the bacteria, viruses, and fungi that are living there. In addition to a bronchoscopy, participants have pulmonary function testing, a CT scan of their lungs, do a couple of six-minute walks, and have blood drawn. All this testing is done without charge to the Alpha and participants will be paid for the time they spend in the study. All these procedure are performed during a single visit to the study site, although it could take up to two days to complete everything.

Although the study has been open for enrollment for several months and we hope to enroll a total of 150 subjects, very few Alpha-1 patients have volunteered for this study. We believe that this is primarily because most Alphas just don't know about it. Also, the fact that a bronchoscopy is included may be a deterrent to some. If you'd like to learn more about this important study or discuss the possibility of participation, please contact one of the following study centers:

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Community Calendars

Alpha-1 Association Education Days

co-sponsored by the Alpha-1 Foundation

February 22
San Francisco, CA

March 22
Dallas, TX

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.

Alpha-1 Foundation Event Calendar

March 3 – Celtic Connection
Boston, MA

April 14 – Friends for a Cure Golf Tournament
Jacksonville, FL

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. 233.

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CONGRATULATIONS!

AlphaNet would like to acknowledge the following SFS Monthly Sweepstakes Drawing Winners:

June 2013:
James Murphy
Phoenix, AZ

July 2013:
Earl Hodson
Ephraim, UT

August 2013:
Gregg Dickson
Petoskey, MI

September 2013:
Sharon Workman
Crossville, TN

Story Ideas?

Do you have an interesting story that you think would make a great article for an upcoming issue of *The AlphaNetter*?

If so, please submit any ideas, comments and/or suggestions to Christine Lanser at clanser@alphanet.org.

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The AlphaNetter

AlphaNet Moves into Newly Renovated Office Building in Coral Gables, Florida

AlphaNet officially has a new place to call home! In November 2013, AlphaNet relocated to a 10,000-square-foot office building in tree-lined Coral Gables, Florida—known as the “City Beautiful.” Having completed a total renovation of the space, AlphaNet can now host a variety of meetings in the technologically-furnished conference rooms. Furthermore, AlphaNet now has the ability to send Welcome Packets and educational materials in a timely and more efficient manner thanks to the designated mail room. Because AlphaNet promotes healthy living, we have included an on-site workout room to reinforce our commitment to exercise. Along with these renovations, other notable features of the building include the installation of upgraded Internet, voiceover IP telephone services and audiovisual teleconference capabilities, as well as security monitoring systems.

While AlphaNet occupies the



Exterior View of the Building



AlphaNet's Second Floor Lobby

second floor, the Alpha-1 Foundation and the COPD Foundation are located on the first floor. AlphaNet has extended “free rent” to the Alpha-1 Foundation to allow this organization to fulfill its mission of helping Alphas without the added financial burden of paying for office space. Furthermore, because AlphaNet and the Alpha-1 Foundation are no longer paying rent, more revenue can be diverted toward research and funding for a cure.

The purchase of the office building serves as a great stepping stone for the future of AlphaNet, as we continually strive to work toward helping Alphas lead better, healthier lives through our disease management programs and clinical research endeavors. With that in mind, we are confident that this building represents the promise and potential for growth in the years to come. Please note the new mailing address: 3300 Ponce de Leon Boulevard Coral Gables, Florida 33134

Winter Weather Tips for Alphas

By: **Bonnie S. Boyd, BS, RN, Director of Disease Management** and **Teresa Kitchen, BSN, RN, Clinical Nurse Manager**

As the temperature plunges outside, it is especially important for Alphas to protect themselves from the elements in order to stay healthy. Here are some helpful tips to heed during the cold winter months:

Wear a scarf or cold weather mask

Cold weather is very drying and can irritate your airways. Take the extra time to gather a scarf or mask before leaving your home. When you have this layer loosely over your mouth/nose area, it can warm and humidify your air, decreasing shortness of breath related to the “icy” cold air.

Exercise indoor versus outdoor

When the air outdoors falls into the single digits and even lower, and you still want to exercise, please remember to warm up prior to your walk. You should spend 15-20 minutes warming up indoors and wear multiple layers when leaving the house. There are also many exercises you can still do inside while watching the snow fall outside.

Avoid fireplaces or wood burning stoves

Alphas are more susceptible to smoke-related problems. Both types of wood burning can expose Alphas to irritation in their airways.

Don't overexert yourself

If you must go outside, work slowly and be aware of the wind chill factor. Ask friends/family to assist with the clearing of the snow/ice from driveways and walkways. Place sand or kitty litter on these areas to increase your footing. Being outside for too long in cold weather can lead to severe shortness of breath.

Hand washing

Frequent and thorough hand washing, specifically after touching surfaces used by others who have been coughing or sneezing, can greatly reduce the potential for infection during this flu and cold season.

Ill family members

Remember that you may need to limit your contact with family members who have a known respiratory illness, especially young children/grandchildren who attend daycare. As we all know, those children are at higher risk for spreading illness.

With these tips in mind, you will be better prepared for what lies ahead in the upcoming chilly months. Here's to staying warm and maintaining good health this winter!

Alpha Motorcyclist Rides for the Cause

Ken Ellis isn't letting his Alpha-1 stand in the way of his love of riding motorcycles. In fact, Ken is doing more than just taking leisurely spins around town; he is making cross-country treks around the U.S. and between the Canadian and Mexican borders to raise money for Alpha-1 and COPD research.

Ken was diagnosed with Alpha-1 Antitrypsin Deficiency over 7 years ago. He had never experienced any significant breathing problems in the past other than what were thought to be some minor asthma-related events when he was a child. This all changed, however, quite suddenly in 2006 when Ken started having more frequent episodes of prolonged breathlessness.

Ken, who was working as a rancher in Idaho, describes one of the first occasions when he knew something was wrong. “I walked into the barn to get some bales of hay for a friend and by the time I had walked back to her with the two bales in hand, I was completely out of breath.” While he had been experiencing some shortness of breath in the weeks prior to this incident, this was really the true eye-opening moment for him.

Shortly thereafter, Ken visited his doctor, who recommended that he see an allergist. The allergist confirmed he had several allergies but also sent for some blood work. These blood tests confirmed that Ken had Alpha-1. With that knowledge, Ken went to a pulmonologist recommended by the allergist and began augmentation therapy soon after.

Ken, who developed his love of motorcycles at the ripe age of ten, decided three years after being diagnosed with Alpha-1 that he wanted to do a cross-country motorcycle ride. “My daughter was graduating from college in 2009 so my wife Cindy and I decided to make a trip out of it. We did a fun ride from Idaho to New Hampshire.”

Following that trip, Ken sought out the next long ride he wished to partake in. “I looked into the Iron Butt Association — a motorcycle endurance riding organization — and decided to do a ride called the “Saddle Sore,” where you cover 1000 miles in less than 24 hours.” Ken raised money for this ride and gave the donations to the COPD Foundation.



Alpha Ken Ellis' view of the road from his motorcycle.

During the same time as the Saddle Sore ride, Ken's mother was going to be celebrating her 70th birthday in Pensacola, Florida so he decided to extend his Saddle Sore trip to surprise his mom at her birthday party. “I tricked my mom a bit by telling her that I was going to be doing a run up North but wouldn't have cell service — which was true.” Ken left Colfax, Washington at 5 a.m. and was in Murdo, South Dakota by 11 p.m. that evening. He had covered just over 1100 miles. “From there, it was a pleasure trip to Pensacola. I left on a Tuesday and by Friday I was having lunch with my mom in Florida!”

Ken's next endurance ride came in July 2013 and was also a fundraiser for the COPD Foundation. The “Border to Border” ride started at the Canadian border and ended past the Mexican border. “Our group started on a Monday morning at 5:15 a.m. on July 22nd and by Tuesday, July 23rd at 2:15 p.m., we had crossed the Mexican border.”

For Ken, his Alpha-1 does not significantly impact his riding and, if anything, he makes minor adjustments to ensure he stays warm and dry. “I am really well protected from the weather. I have a helmet — with my Bluetooth attached — that shields me from the elements and I also have an adjustable windshield, which I can move up and down as the weather dictates. What I find helpful, too, is the portable nebulizer that I bring which has a 12-volt plug and has its own battery.” All these tools make for a safer and easier ride for Ken.

Ken is already looking forward to his next ride, which he is currently organizing for July 2014. This ride, known as “Heaven to Hell,” commences at Pikes Peak in Colorado, where the highest elevation of paved road in the country is located, and ends in Death Valley, California, where the lowest elevation of paved road in the US is found. This ride will raise donations to benefit the Alpha-1 Foundation.

It is evident from his passion for riding and his love of the open road that Ken won't be hanging up his helmet anytime soon!

Clinical Corner

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

Are you compliant with your medication? Have you or a family member ever asked, “I can't really tell if this medicine is helping me, so why do I bother taking it?” Taking medicines as ordered by your physician is of the utmost importance.

Here are the most common reasons people are non-compliant with taking their medicines:

- *Fear of addiction
- *Fear of side effects/experiencing side effects
- *Overwhelmed with the number of pills or inhalers prescribed
- *Skipping doses to save money
- *Difficulty opening pill containers
- *Forgetfulness about taking meds

Remember, even if your symptoms disappear, you need to continue taking the medication for the full course ordered by your doctor. Studies have shown that with proper medicine compliance, patients can avoid having their condition worsen and they will also have a better quality of life.

Here are some helpful hints to keep you on track with taking your medications:

- *Set up a reminder by using a pill container, an alarm or a daily journal
- *Get into a routine by taking pills when you first wake up, with meals or with other set activities
- *Work as a team with your caregiver, spouse and/or other family members by explaining to them why you take each medication and the purpose (i.e. blood pressure pill, inhaler, sleeping pill, etc).

Step Forward Study Updates

- We are at the midway point of the last year of the 5-year-long clinical trial, with the conclusion being August 2014.
- The Daily Diary Sweepstakes is on-going, so we encourage you to continue entering your data either online or using the mail-in postcards in order to qualify for the monthly drawing.
- Please remember to send back your AlphaNet flash drive in the prepaid envelope provided to you, so that we can gather your updated spirometry data.
- If you have any SFS-related questions, please contact Communications Specialist Ashley Chase at achase@alphanet.org.