

AlphaNet Coordinator Shares Smoking Cessation Story

The importance of smoking cessation for Alphas cannot be stressed enough. The Big Fat Reference Guide (BFRG) notes that, “for Alphas, smoking is the greatest risk to your health, leading to the development of lung disease and premature death. A single cigarette destroys all the alpha-1 antitrypsin in the lungs.” Gayle Allison Tipper, AlphaNet Coordinator since 1999, has agreed to tell her story of how she quit smoking:

“I was a smoker for about 14 years. I had stopped smoking 6 months before being diagnosed with Alpha-1. When I was 32, I had an upcoming foot surgery and, during that time, I had bronchitis and couldn’t quite get over it. Just to be safe, the surgeon ordered a chest x-ray. The chest x-ray came back indicating severe COPD, so we had to see why someone so young, who appeared to be healthy, had such bad lungs.

I went home and looked up COPD in a Mayo Clinic reference guide dated 1981 (I didn’t have a computer then), and in one paragraph under Emphysema it stated: “In rare instances a deficiency of alpha-1 antitrypsin can cause early onset emphysema in adults ages 30 to 50.” I wrote down the name and took it to my PCP, who tested me, and sure enough I was a ZZ, FEV1 was 35% and they told me my lungs were 118 years old.

I had stopped smoking 6 months earlier, and my pattern was to stop smoking and then, when something stressful came up, I would start smoking again. This time, I was fortunate that the diagnosis made it easy not to restart smoking. However, it took me many attempts to finally quit.

I was not the typical smoker who had to have a cigarette as soon as their feet hit the floor. I would have my coffee first and then head to work. Usually, I had my first cigarette around 10 a.m. during a break at work, and then continued to smoke about 1 pack a day. I usually had my cigarette as my “dessert” when others had the real thing. I was a petite size 5 and had no clue what I was doing to myself. When I finally wanted to stop smoking for myself, as well as for my children, I tried the patches but they didn’t seem to work for me. So, I tried the nicotine gum. The first few times it didn’t seem to work and I would give up after about two weeks. But then someone told me to put something in my hand to “fiddle” with. So, I cut a long straw in two, and I would hold on to that. I even inhaled air through the straw to get the effect of smoking while I packed the gum in my jaw when I had a bad craving for a cigarette. The other thing that helped me was I usually drank my coffee with cream and sugar, but changed to black coffee when I wanted a cigarette really badly and it kind of “shocked” my system so I didn’t want either of them. As strange as it sounds, this is what really worked for me. The desire to quit, the gum, straws and black coffee.

It is now year 17 since I was diagnosed with Alpha-1. I recall AlphaNet Medical Director Dr. Robert A. Sandhaus saying that smoking can take 10 years off your life, and I can honestly say that the past 13 years have been the best years of my life. I sure am glad I didn’t take those 10 years off! I have been blessed with the best job you could ever have as an AlphaNet Coordinator for almost 15 years, married the man of my dreams 5 years ago, and have been a foster parent for the past two years. I plan on the next 20 being even better. Life is good!”

For more information on smoking cessation programs, please contact your AlphaNet Coordinator or consult the BFRG: <http://www.alphanetbfg.org>.

Community Calendars

Alpha-1 Foundation Education Days

November 1
St. Louis, MO

December 13
Las Vegas, NV

To find out more about conferences and support groups, visit the Alpha-1 Foundation website at www.alpha1.org or call toll-free: (888) 825-7421, ext. 331.

Alpha-1 Foundation Events Calendar

Nov. 8 – Step Forward for Alpha-1 Walk
Myrtle Beach, SC

Nov. 15 – Step Forward for Alpha-1 Walk
West Palm Beach, FL

Dec. 11 – Las Vegas Rocks Alpha-1
Las Vegas, NV

To find out more about these or other events in your area, log on to www.alpha1.org or call toll-free: (888) 825-7421, ext. 233.

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For Your Information...

Your AlphaNet Coordinator is a great resource, whether you are a newly diagnosed Alpha or a long-time AlphaNet subscriber. Take advantage of his/her expertise.

Check out the “**Questions to Ask Your Coordinator**” pamphlet and other FAQ’s at www.alphanet.org.

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’s Step Forward Study Ends with a Bang

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

A five-year-long study, designed to evaluate a variety of ways to improve the health of individuals with Alpha-1, has just reached its end. A total of 500 subjects consented to join the study, known as the Step Forward Study (SFS).

The motivation for this study began when we looked at the data from the original Outcome Study started in 2003. The Outcome Study looked at the potential benefits of ADMAPP (the AlphaNet Disease Management and Prevention Program) by following 1,000 individuals with lung disease due to Alpha-1 before and after starting the ADMAPP program. It found that ADMAPP had positive effects on many aspects of its participants’ lives. It reduced exacerbations, ER visits, antibiotic usage, steroid usage, and improved overall quality of life.

However, ADMAPP did not seem to improve a participant’s exercise performance, and we found that most participants were far from their ideal weight because they weighed too much or too little.

The SFS study was designed to find out what interventions AlphaNet could perform that would improve exercise and nutrition. After enrollment, participating Alphas were randomly assigned to two different groups of 250 each. One group received the usual, excellent care that all individuals enrolled in the various AlphaNet programs receive (the control group). The other group received the same care plus some innovative additional interventions designed to improve their exercise and nutrition (the treated group). Now that the study is over, we can reveal some of what happened with each of these groups!

During the course of the study, we introduced new interventions approximately every six months. Since we know that our AlphaNet participants talk to each other at support groups, in blogs, on Facebook, by phone, and by email, we had to find a way to interact with the two groups that would minimize the likelihood that they could learn which group they were assigned to. We did this by rolling out something to people in both groups at the same time as we rolled out an intervention to the treated group. When the treated group received its first intervention (Therabands and an exercise poster), both groups

were sent the Skinny Little Reference Guide (SLRG) entitled, *Exercise for Individuals with Lung Disease*. Other items we sent to everyone included AlphaNet water bottles, the AlphaNet Healthy Cookbook, and a harmonica with a guide on how to use it to improve breathing. In addition, the treated group also received mini exercise bikes (peddlers), an exercise ball with a poster for its use, instructions for using canned goods as weights for strength improvement, and teleconference interventions by a nutritionist. The final and most dramatic intervention was a DVD disc set, teleconferences, and written materials for doing pulmonary rehabilitation at home. We called this Virtual Pulmonary Rehab or VPR.

People who participated in the SFS also were given home spirometers to check their respiratory status twice a month and sent flash drive memory cards back to AlphaNet with their spirometry data so it could be analyzed as part of the study. Finally, we made a variety of data collection options available to the SFS participants including sending data by postcard, by personal internet sites, and, toward the end of the study, by a proprietary iPad app.

By the end of five years, the AlphaNet database has collected close to 3 million meal entries, over 100,000 weight entries, nearly 50,000 spirometry tests, about 24,000 monthly questionnaires, and approximately 2,000 quality of life questionnaires. As you can imagine, it will take some time to evaluate and analyze all the data collected during the study. But these numbers do point to the most important aspect of the SFS program: the amazing effort put forth by our Alphas and our Coordinators to accomplish this task. Thank you, thank you, thank you! The SFS participants are the true heroes of this story. The medical team thanks you for your incredible effort.

In the coming months, SFS participants will be provided with updates on the course of our analysis and our expectation is that several scientific publications will result from this study. In the end, thanks to the efforts of all those mentioned, we believe this study will improve the quality of life of Alphas around the world.

Clinical Trials in Alpha-1 Antitrypsin Deficiency

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

Many of our AlphaNetters have participated in clinical trials over the years. In fact, it's thanks to the clinical study participation of our AlphaNet group that we now have four intravenous augmentation therapy products in the U.S. instead of just one! Many people with Alpha-1 feel they owe it to their children and grandchildren to help find the best treatments for this disease and, someday, find the cure.

But not every study is on a direct path to the cure for Alpha-1. And most studies have risks associated with participation that need to be weighed against the potential benefits. Some studies are designed to provide information about the diseases associated with Alpha-1 rather than test a new therapy. Some tests of new therapies involve real potential risks, often because they are employing drugs or delivery methods that have rarely been used in humans.

What happens if you enroll in a study and you have a bad reaction to the new medication or a medical procedure? Questions about such risks and the potential benefits of a given trial can be found by carefully reading the consent form that all studies must provide and by discussing these questions with the investigator or the study staff. Never sign a consent form without reading and understanding what it says.

You might be surprised how many studies involving Alpha-1 are going on currently or are planned to start within the next several months. Because people frequently call their AlphaNet Coordinators to ask about the studies currently enrolling, we thought it might be a good idea to list some that you know about and some you may not know about. Most of the information here is available by searching the U.S. government website: www.clinicaltrials.gov.

Study Name: Efficacy and Safety of Alpha-1-Proteinase Inhibitor (Human), Modified Process (Alpha-1 MP) in Subjects with Pulmonary Emphysema Due to Alpha-1 Antitrypsin Deficiency
Nickname: SPARTA **Sponsor:** Grifols **Sites:** Multiple U.S. Sites
Description: Comparison of 60 mg/kg/week intravenous dosing with 120 mg/kg/week and placebo. Total of 100 patients per group and each patient randomized to one of the three groups for three years of treatment.

Study Name: Genomic Research in Alpha-1 Antitrypsin Deficiency
Nickname: GRADS Alpha-1
Sponsor: National Heart, Lung, and Blood Institute of NIH
Sites: Multiple sites in U.S.
Description: One-visit study enrolling individuals with ZZ (on and off augmentation therapy) and MZ (off augmentation therapy). Visit includes questionnaires, PFT, CT of chest, blood work, and a bronchoscopic washing of the lungs. Goal is to learn whether the genetics of the viruses, bacteria, fungi, and mycobacteria living in the lungs affects disease severity.

Study Name: Adeno-associated Virus Delivery of Alpha-1 Antitrypsin Gene Therapy by Isolated Limb Infusion
Sponsor: AGTC and TAP
Sites: National Jewish in Denver, University of Cincinnati, and University of Massachusetts.
Description: Gene therapy trials to enroll three subjects in each of two dose groups. Subjects will be transported to a facility with experience in isolated limb infusion (currently Duke University) and will have one leg delivered gene therapy under general anesthesia. Subjects will have to commit to be off all augmentation therapy for a minimum of one year.

Study Name: Effect of Double Dose of Alpha-1 Antitrypsin Augmentation Therapy on Lung Inflammation
Sponsor: Grifols
Sites: Dr. Michael Campos, University of Miami
Description: Evaluating whether 120 mg/kg/week improves lung inflammation.

Study Name: Alpha-1 Antitrypsin Deficiency Adult Liver Study
Nickname: A-1F liver study **Sponsor:** Alpha-1 Foundation
Sites: Multiple sites in U.S.
Description: Study designed to evaluate the natural history of liver disease in Alpha-1 adults. Individuals will expect to undergo usual liver function evaluations plus liver biopsies on at least 2 occasions.

Study Name: Alpha-1 Antitrypsin Deficiency Adult Liver Study
Nickname: Dr. Mark Brantly's liver study
Sites: University of Florida
Description: Liver biopsy study.

Study Name: Phase II, Safety and ELF Study of "Kamada-API for Inhalation"
Nickname: Kamada inhaled U.S.
Sponsor: Kamada, Israel
Sites: University of Florida/ University of Texas at Tyler
Description: Study of inhaled alpha-1 antitrypsin (Glassia) to evaluate the safety of chronic inhalation in the U.S.

Study Name: Carbamazepine in Severe Liver Disease Due to Alpha-1 Antitrypsin Deficiency (CBZ)
Nickname: Severe liver study using Tegretol
Sponsor: University of Pittsburgh
Sites: Dr. David H. Perlmutter at University of Pittsburgh
Description: Trial to see if the existing drug, Tegretol (carbamazepine), can reverse the changes of severe liver disease.

Study Name: Safety Study of Gene Transfer Vector to Treat Alpha-1 Antitrypsin Deficiency
Sites: Dr. Ron Crystal, Cornell Medical Center, NYC
Description: Not yet enrolling but expected to be an early safety study of a novel delivery method for gene therapy in Alpha-1. Plans to deliver the gene therapy to the line membrane of the lungs.

In addition to the studies featured, there are a lot of other exciting studies on the horizon. Most are looking at drugs to prevent or treat liver disease. We'll try to let you know of new studies as they start recruiting.

AlphaNet Welcomes New Coordinators



Ernie Ingles

Ernie Ingles joins AlphaNet Canada after a 42-year academic career. From 1990 to 2014, Ingles served as Vice-Provost at the University of Alberta, where he was responsible for the Library System, the Department of Museums and Collections, the University of Alberta Press and other duties. Previously, he also served as the University's Chief Information Officer, as well as the Executive Professor and Director of the School of Library and Information Studies.

Ingles has been active within library and information technology communities, having served in numerous professional associations in senior executive and board positions.



Stephen James

Stephen James joins AlphaNet as a Zemaira Coordinator after a 24-year career at AT&T, where he developed budgets and reported on the monthly performance of in-bound call centers. Following his diagnosis, James became passionate about educating himself and others about Alpha-1. He attended his first National Conference in 2000.

In 1998, he participated in a research study conducted by Dr. Mark Brantly, and began attending Alpha-1 fundraisers and events. Although James considers the Alpha community a diverse family, he realizes that Alpha-1 serves as a common thread that brings everyone together.



Lois Lange

Prior to joining AlphaNet as a Zemaira Coordinator, Lois Lange served as an Alpha-1 mentor for AccessMed and is currently on the Board of Directors for the Idaho Alpha-1 Community Outreach, Inc. support group.

Raised in Bruneau, Idaho, a small farming and ranching community with a population of 500, Lange was diagnosed in 2006 with Alpha-1. Her husband, Jerry, was diagnosed three years earlier. Both Lange and her husband are on augmentation therapy. Lange's personal experience living with Alpha-1 has motivated her to help educate and support others in the Alpha-1 community.



Terri Leffler

Terri Leffler joins AlphaNet as a Baxter Coordinator.

Her career has taken her down many paths, from business owner to a journey in sales and marketing — which included positions in the communications and medical field. Leffler's life quickly took a sharp turn in 2006 when she developed severe lung infections and shortness of breath.

In 2010, Leffler discovered that her home was full of several kinds of toxic mold, which had been causing her prior health issues.

As a Coordinator, Leffler encourages Alphas to make positive changes in their lives, such as eating healthy, staying positive and doing something you love every day.



Stephen Petty

Stephen Petty joins AlphaNet as a Baxter Coordinator

with over 30 years of management experience. Petty spent 14 years working with a pharmacy chain before going into business on his own, designing and remodeling independent pharmacies in the Northeast.

Petty was diagnosed with Alpha-1 in October 2005, after switching to a new pulmonologist. In December 2005, Petty started augmentation therapy and spent the next seven years in the transplant program.

Prior to AlphaNet, Petty served as the Support Group leader of the Connecticut "Nutmeggers."



Diana Sisler

Diana Sisler joins the AlphaNet Baxter Coordinator

team with a professional background in healthcare. One area of Sisler's concentration was increasing clinical professionals' education of the signs and symptoms of Alpha-1, as well as increasing awareness of testing. It was during this time, and also in the midst of an unusually rough allergy season, that Sisler's colleague suggested that she take the mail-in blood test for Alpha-1. Soon after being diagnosed, she began augmentation therapy and has been infusing for over two years.

Sisler is excited to be a part of AlphaNet and hopes to become a trusted, caring resource for those with Alpha-1.

Clinical Corner

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



It's Time to Discuss Vaccines!

It is hard to believe that Fall is upon us and that it is, once again, time to remind Alphas of the upcoming flu season! Here is some valuable information about the flu vaccine, as well as info about other vaccines too.

Flu Vaccine

- Annual flu shots/vaccines are recommended for Alphas. It is important to remember that the nasal spray vaccine (FluMist®) is a LIVE vaccine, as it contains a live virus, and if you are a transplant patient (liver or lung), you should NOT receive it, but receive a flu shot. Please remember that it is important to remind your family and people you live with to be vaccinated.

Pneumonia Vaccine

- It is recommended to have Prevnar 13 (also called PCV 13) every 5 years. The older version of the vaccine is called Pneumovax. AlphaNet Medical Director Dr. Robert A. Sandhaus explained that there is no specific recommendation about whether everyone who has had a Pneumovax injection less than 5 years ago should receive the PVC13 vaccination sooner than 5 years after the Pneumovax. However, it was recommended that if you have received Pneumovax in the recent past, you shouldn't receive PCV 13 sooner than one year after the Pneumovax. If you are a transplant patient, it would be best to contact your physician.

Shingles Vaccine

- This vaccine is recommended for all adults over the age of 50 or any adult who has had a shingles infection in the past. Because this is a LIVE virus, it should NOT be given to an individual who is immunosuppressed, such as people who have had a lung or liver transplant. In fact, family members of transplant recipients should speak to their doctors before being immunized as they can infect their transplanted relative if they receive the vaccine.

Whooping Cough (Pertussis) Vaccine

- Most of us were vaccinated against whooping cough or pertussis when we were children. Alphas were encouraged to receive a booster of the vaccine prior to attending the Seattle Alpha-1 Association National Conference a few years ago. There is a recommendation that all adults 65 years and older be re-vaccinated for pertussis. You only need this vaccination once as an adult, but as an Alpha your physician may recommend it if you are younger than 65.

If you have any further questions about vaccines, please don't hesitate to call your AlphaNet Coordinator or search the www.cdc.gov site for additional information.