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The AlphaNetter is a publication of AlphaNet, Inc.

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# The AlphaNetter

## AlphaNet Contributes \$225,000 to the John W. Walsh Translational Research Award

During this year's 17<sup>th</sup> annual Alpha-1 Foundation Celebration of Life, AlphaNet announced that they would be donating \$225,000 to the John W. Walsh Translational Research Award. This contribution matches the \$225,000 gift from Mark Brantly, MD and his wife, Judy Lew, MD, who established the John W. Walsh Translational Research Award. These donations are part of the larger John W. Walsh Research Fund, which was created to advance the scientific and clinical understanding of Alpha-1 Antitrypsin Deficiency to help find the cure.

"AlphaNet's matching \$225,000 gift to the John W. Walsh Translational Research Award honors John's legacy, as he dedicated his life to finding a cure for Alpha-1 and focused his energy toward improving the quality of life for every Alpha. This award will propel his mission forward by encouraging therapy-oriented translational research with the goal of turning research findings into practical medicine," AlphaNet President and CEO Robert C. Barrett said.

## AlphaNet Donates Funds for Educational Scholarships

AlphaNet recently donated \$10,000 to the Alpha-1 Foundation to fund four \$2,500 educational scholarships for deserving Alpha-1 affected individuals. The scholarship recipients, who are either Alphas or immediately related to an Alpha, were selected by the Alpha-1 Foundation for their involvement in the Alpha-1 community and their continued dedication to the cause. Each scholarship honors the memory of an influential individual in the Alpha community: 1.) John (Jack) W. Walsh III, father to John, Fred, Judy and Sue, who emphasized the importance of education and family within his immediate family and within the entire Alpha-1 community; 2.) Robert (Bob) Haggerty who was the face and voice of the Alpha-1 Association for almost a decade and served as Treasurer for many years, as well as Master of Ceremonies at the National Conference; 3.) Terry Young who served as the first AlphaNet Coordinator and later as AlphaNet General Manager and was a mentor to all AlphaNet Coordinators and staff members; 4.) Emma Louise (Lou) Carr Glenn who was an exceptional member of the Alpha-1 Association Board of Directors for nine years, and the loving mother and caregiver of AlphaNet Coordinator Patti Brown.

# AlphaNetters Want to Know: What Has AlphaNet Learned from All the Questions Our Coordinators Ask Us Each Month?

By: **Dr. Robert A. Sandhaus, MD, PhD, FCCP, AN Medical Director**

Most AlphaNetters know that AlphaNet employs, trains, and supports their wonderful AlphaNet Coordinators (the ANCs). They know that we created the BFRG (the Big Fat Reference Guide to Alpha-1) and the 13 SLRGs (Skinny Little Reference Guides) covering a bunch of important topics. They may even know that AlphaNet has donated over \$50 million to the Alpha-1 Foundation to support its research mission in quest of curing Alpha-1 Antitrypsin Deficiency. And that the total population of individuals followed by AlphaNet has grown from about 100 in 1998 to over 6,000 participants today.

Some of you may not realize that AlphaNet has been a research powerhouse of its own, ever since its founding in 1995. There is not an augmentation therapy in use today that AlphaNet didn't have a role in studies leading to their approval. From the original studies leading to the marketing of Aralast, Zemaira, and Glassia, to the study that led to the approval of the change from Prolastin to Prolastin-C, AlphaNet had a role in identifying study sites and recruiting patients. Sometimes we organized and educated the nursing services involved in blinding and administering drug. In almost all studies AlphaNet acted as the travel agent, arranging flights and hotels and taking care of reimbursements (thank you, Kathie Sivesind!).

More than that, AlphaNet has funded its very own research projects over the years. You may have noticed that each month when an ANC calls you on the phone, in addition to answering your questions and finding out the latest on your health, they ask you a slew of questions. Those of you who've been in the AlphaNet system for years may know them by heart. Sometimes the answers don't change from month to month or year to year – even the ANCs get bored. Other times they chart a meandering course of someone with frequent flares of their lung disease, including admissions to the hospital or even a lung or liver transplant.

So, what happens to all these answers you provide? In fact, those answers, now accumulating from over 6,000 AlphaNet participants each month, make up a collection of information that has allowed us to evaluate various treatments, provide information about the likelihood that a patient with Alpha-1 will find themselves in a hospital each year, and has led to over 30 publications in medical literature.

There are several commitments that AlphaNet has made to you. We never provide or submit any identifying information in any publication. We don't share your identifying information with any company we work with or investigator who asks to learn about our patients. We often do research with renowned investigators at institutions around the world, but AlphaNet always maintains strict controls on the information that you have provided to us.

Let me tell you a little about what we've learned in evaluating all the answers you've provided to our questions. In 2005, two papers were published, based on information that you provided to AlphaNet, which showed that in spite of all the information that the Alpha-1 Foundation and others have provided to doctors and other healthcare professionals, there have been no major improvements in detection of people with Alpha-1 compared to a similar study from the 1990s. The only positive take-away was that more older individuals were being detected; suggesting that our message was getting through that everyone should be tested regardless of age or smoking history.

In 2008, we published the information that there are four major risk factors for the development of lung disease in Alpha-1: smoking, secondhand smoke, occupational exposures, and frequent lung infections.

In 2009, we published one of our first AlphaNet papers, written with Dr. Michael Campos and the pulmonary group at the University of Miami. We found that, on average, people followed by AlphaNet had 2.4 exacerbations per year and each exacerbation lasted an average of 17 days. The more exacerbations a person had each year, the worse his/her quality of life was and, as you might expect, those with more exacerbations had more frequent doctor visits, emergency room visits, and hospitalizations.

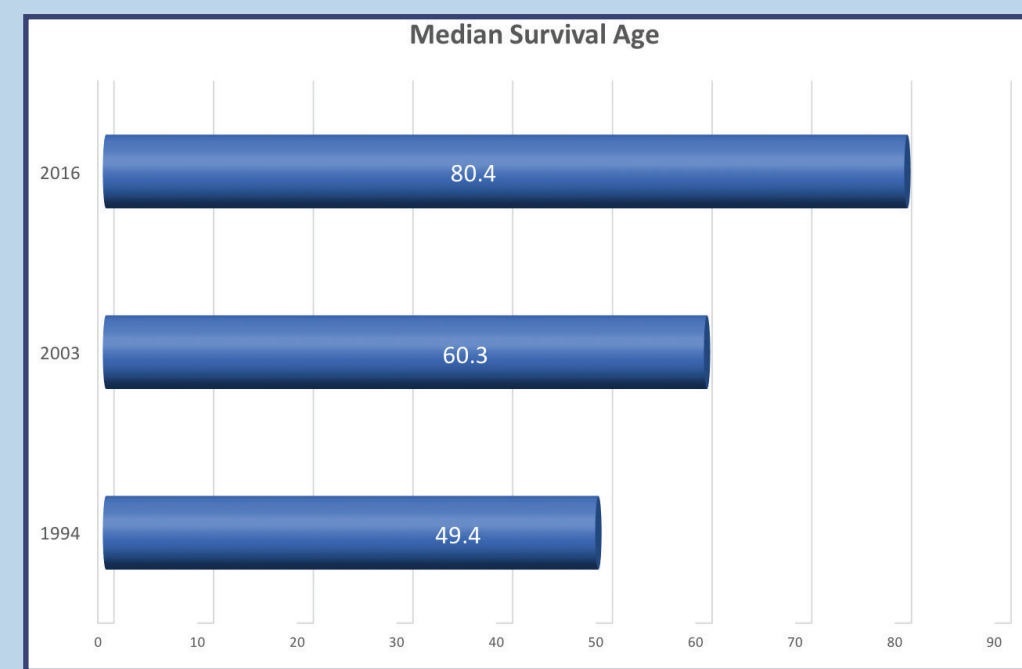
Most importantly, in 2009 we published the results of AlphaNet's Outcome Study comparing the health of AlphaNetters before and after the start of our AlphaNet Disease Management and Prevention program or ADMAP. We followed 1,000 AlphaNet participants over two years and showed that after participating in ADMAP for a year, there were significant drops in medication use, exacerbations, emergency room visits, unscheduled physician visits, and days spent in the hospital. These were all much more dramatic changes than we expected to see in only a single year of ADMAP. But all was not perfect. In spite of a concentration on improving exercise and nutrition, the ADMAP study did not show any improvement in these outcomes.

Based on these results, AlphaNet decided to embark on a five-year study to try to improve exercise, nutrition, and weight in our participants. This was the origin of the Step Forward Study (SFS), a study designed to compare 250 AlphaNet participants on our usual ADMAP program with 250 participants selected at random to receive a program we call Virtual Pulmonary Rehabilitation (VPR). The VPR group received exercise and nutrition aids, personal intervention through teleconferences, DVDs with exercise and daily living suggestions, and nutritional education.

The results of the SFS study have finally been compiled and are being prepared for publication. We have always promised our AlphaNet participants that they would be the first to learn about the results of the studies that AlphaNet performed, and that is one reason for this article. While we are still analyzing the data, some important results have already been found – some of which were presented in posters at the American Thoracic Society meeting in May of this year.

Participants in SFS were very diligent in completing the study and answering the monthly questionnaires, doing their twice monthly spirometries, recording their weight, food intake, pedometer readings, and exercise. The participants in the VPR group had statistically significant increases in exercise time and walk distances compared with the control group. People in the VPR group moved towards their ideal weight goals dramatically more effectively than those in the control group (those who were underweight gained weight, those who were overweight lost weight). We've started to analyze the spirometry (lung function) data, but we have about 100,000 pulmonary function tests to evaluate, so that work is moving a bit more slowly.

But the most significant results of all relate to the survival of people within the AlphaNet programs. In 1994, a publication from Europe looking at Alpha-1 patients in a country where augmentation therapy was not available, showed that the median survival (some call it average life expectancy) of an Alpha with lung disease was about 49 years of age. This year, AlphaNet looked at our participant population and found that Alphas on augmentation therapy – but before starting the ADMAP program – have a median survival of over 60 years. And when ADMAP is added to augmentation therapy in people with Alpha-1 lung disease, the median survival rises to over 80 years of age (**see figure**).



Another thing the Coordinators do during their monthly calls with AlphaNetters is find the answers to questions that can be answered with one simple set of questions. Some recent examples include a set of questions that was asked once regarding the presence of depression and other psychiatric conditions in families with Alpha-1. An additional set of one-time questions was also added about macular degeneration (a disease of the retina of the eye that can cause blindness) among Alphas. In short order, we found that the rates of psychiatric problems and macular degeneration were no different than in the general population. Similarly, we've recently asked how many people on augmentation therapy self-infuse and how many might want to learn. Please expect future one-time questions about premature labor in Alphas, eosinophilic esophagitis in Alphas, and lung cancer in Alphas. In addition, the Speck Study will be starting, looking to learn if high particulates in the air are associated with more frequent lung infections. This will be done by placing a free particulate monitor in the homes of those who want one and who qualify to participate.

It is worth pointing out that the AlphaNet population does not represent the entire population of people with Alpha-1. Because most patients learn about AlphaNet when they are started on augmentation therapy, our population is heavily weighted towards Alphas with lung disease being treated with augmentation therapy. Over 90% of our AlphaNetters fit this profile. This will likely change somewhat as more people join AlphaNet's REACH program, which is designed to follow healthy individuals with Alpha-1 and Alphas with liver disease. Still, the vast majority of people living with Alpha-1 remain undiagnosed.

There are many more questions to be answered, both in terms of learning as much as possible about Alpha-1 and its effects on individuals and in terms of the calls you receive each month. ***I hope this helps people understand why these questions keep being asked – over and over and over – and I hope you will continue to answer these questions.*** I guarantee your answers help the Alpha-1 community and shed new light on lung and liver diseases in general. ***To our AlphaNet participants: Thank you all for the time and effort you spend answering these questions each month.***



# Clinical Corner

By: **Teresa A. Kitchen, BSN, RN, AN Clinical Nurse Manager**

As Summer winds down and Fall is fast approaching, it is a good time to discuss what individuals with Alpha-1 can do to minimize their risk of acquiring respiratory infections, as well as avoid certain environmental risk factors. As outlined in AlphaNet's Skinny Little Reference Guide entitled "*Staying Healthy: Management of Environmental Risk Factors*" we learn what risk factors Alphas should be aware of and avoid, if possible.

First, we all know that smoking is the greatest risk associated with the development of lung disease in Alpha-1 individuals. Secondhand smoke can also cause damage to the lungs of Alphas who do not smoke themselves. A single cigarette destroys all the alpha-1 antitrypsin in the lungs. So, although smoking cessation may be a difficult lifestyle change to make, it is the most important step you can take to improve and preserve your quality of life.

Second, there are many risk factors in the home, work and outdoors. Household cleaners, bug sprays, fireplace smoke, fertilizers and fumes can be easy triggers to cause breathing difficulty for Alphas. Make sure to have ventilation in areas where these fumes are found and wear a mask as necessary. You may also want to consider wearing a mask when working outside in the yard. Pollen and fertilizers can be quite irritating for people who have seasonal allergies, as well as those with Alpha-1. While no mask can provide 100 percent protection, N-95 respirators are a specialized type of disposable particulate respirator. Simple earloop masks do not prevent the wearer from acquiring airborne infections from others. Always look to see if the mask has been NIOSH approved (National Institute for Occupational Safety and Health).

Third, the importance of good hand washing technique is crucial to avoid the spread of infection. Remember, you don't need to use expensive antibacterial soap; the important take-away is the benefit of using soap combined with the friction of rubbing your hands together and proper rinsing. Avoid putting your hands near your mouth, as this is a common way that individuals can spread illness.

Finally, vaccines are also important to decrease your risk of serious complications associated with certain diseases. Discuss with your doctor the proper time to receive the flu vaccine, pneumonia vaccine and hepatitis A and B vaccines.



## AlphaNet Appoints New Board Member



Dave Pusey was elected to the AlphaNet Board of Directors this past February. Dave is the husband of the late Barb Pusey, who was the first AlphaNet CSL Zemaيرا Coordinator and later served as the Zemaيرا Program Manager.

Dave worked for 35 years as an engineer/manager for Eastman Kodak in facilities design and construction. He also worked for the City of Fort Collins Utility as an environmental engineer and for Carestream Health as the Environment, Health and Safety Manager. Dave has served on the Fort Collins American Cancer Society Board in several capacities including Chair. He also served two terms on the Colorado Water Quality Control Commission as member and Chair. He is currently a member (past Chair and Vice-Chair) of his local Fire Protection District and the Poudre Fire Authority. Dave looks forward to working on the AlphaNet Board to continue Barb's enduring legacy.

# Diagnosis Day

*AlphaNet Coordinator Siobhan Lestina shares her Alpha-1 diagnosis story and how she took control of her diagnosis with the support of her family, friends and medical team.*

Seven years ago on June 30<sup>th</sup>, I received the news that would change my life forever. Little did I know at the time that this would also be the beginning of a new chapter in my life, one that would bring new meaning and, in a way, breathe new life into me.

I remember where I was standing when I got the news – in my bedroom, getting ready to go see the new Twilight saga movie. I had been suffering from various breathing issues previous to this, but put it down to my asthma, to weight gain from having two babies close together, or being worried about living in Cheyenne, Wyoming with a young family and having no support as our families lived many driving/flying hours away. I always had an excuse for what the cause was because I didn't want to believe that it could be anything serious. We were getting ready to move to St. Louis, Missouri to be closer to my husband Ryan's family. He was close to being hired for a new job there and it seemed like things were finally starting to turn around. I remember standing in that bedroom thinking that things were starting to look up and that the worst was finally behind us.

Then came the phone call. I heard the phone ring and I knew. I heard Ryan say "Hi, Dr. Popova, I'll get her," and I knew. I knew if the doctor was calling after hours that the news couldn't be good. I will never forget those words, "You have the deficiency. You are a very sick girl but we are going to help get you better." It was the first time I had been called a very sick girl for something other than my sense of humor!

I will never forget that feeling of hopelessness. That feeling of not knowing if I would see my babies grow up, my babies that we had tried so hard for. We had overcome the odds to have two beautiful, healthy children and now it seemed as if they were going to have their mommy ripped away from them. It seemed like the world's cruelest joke.

I didn't know much, or really anything, about Alpha-1 Antitrypsin Deficiency other than it is what took my Aunt Grace from this world. She died when I was in college and only her children were advised to be tested at that time.

I convinced myself I was dying and started living my life that way. I felt like I didn't have anyone to talk to. We had moved to St. Louis and away from the doctor who had diagnosed me and I didn't know where to go from there. I found a pulmonary specialist who unfortunately did not know how, or feel comfortable treating me and decided not to take my case. Instead of finding another doctor, I fell into a mild depression, but I knew I had to keep it together for my kids, and they helped keep me from completely falling apart.

Ryan was, and still is, my rock. It could have been easy for him to walk away and not have to deal with a chronically ill wife who cried a lot, and on occasion was bitter and angry, but he

didn't. He supported me, even on my darkest days, and he understood my frustrations and accepted my mood swings. He hid his own fear so as not to upset me, and he let me come to terms with my diagnosis in my own time.

My "own time" took almost two years. In 2012, I went to a pulmonary specialist as recommended by my new primary care physician. I walked into her office and her nurse greeted me with a huge smile and asked if I was the new patient with Alpha-1. When I confirmed this, she became excited because her sister worked at the office where the augmentation therapy I would be receiving came from, and she enthusiastically promised me that they were going to take care of me. For the first time in the two years since my diagnosis I genuinely felt that everything could be okay. The doctor admittedly did not know much about

Alpha-1 and I was her first patient, but she was so willing to learn about it that I knew I had come to the right place. Within days, I had a phone call from my patient service center explaining the infusions to me, how they could help, and informing me that there was a patient support organization for Alphas called AlphaNet and would I be interested in having them contact me.

I finally had that huge weight that had taken residence on my shoulder evicted! There was hope again – hope that I would get to see my kids graduate high school, college, get to see them marry, maybe even see my grandkids! And not only that, but there were other Alphas out there, and they wanted to talk to me! I felt like I could breathe again...metaphorically speaking!

It was time to start living again.

And live I did. I started

traveling again and decided that, with

the help of a little O2 I did not have to be restricted. I volunteered for the Parent Teacher Organization (PTO) and tried to go to as many of my kids' school functions as physically (and mentally) possible. I went to the community college to brush up on my computing and Microsoft Office skills, and I even walked a few 5Ks. I have 5K walk scheduled this Christmas.

I attended my first National Conference in 2016 and was so inspired by the people I met and the education that was offered, that I came home and told Ryan that this is what I wanted to do. I wanted to help other Alphas the way I had been helped. Four months later, I started working for AlphaNet and have met the most amazing, encouraging, and inspirational people. The courage, compassion, and love that exudes from them is wonderful and I can see them making a difference in the lives of others, as they did for me five years ago.

I was able to bring Ryan to the conference this year and introduce him to my AlphaNet family. Being a part of this community has been a humbling experience. Seven years ago, I had lost all hope and thought my life was over. Now, seven years later I am reflecting on how this diagnosis has brought me a life that would never have been possible otherwise. I love what I do and I can only hope that I can bring some hope to those who are finding themselves in the same place I was not so long ago.



*Siobhan and her family enjoy Disney World.*

## Alpha-1 Foundation Education Days

September 30  
Boise, Idaho

October 21  
New York/New Jersey

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

## Alpha-1 Foundation Events Calendar

September 22 - 24  
Team Alpha-1 Escape to the Cape  
Plymouth, Massachusetts

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

### 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](http://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 Yllanes at [cyllanes@alphanet.org](mailto:cyllanes@alphanet.org).

