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The AlphaNetter

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ALPHANET WELCOMES 2 NEW COORDINATORS



Jane Totten and Marilyn Wagner have recently joined the AlphaNet family as Coordinators. AlphaNet is pleased to have these two very special women on board to help educate, counsel, and assist others, like themselves, with Alpha-1.

Jane resides in Robbins, North Carolina and is providing support services to Alphas in Georgia, South Carolina and Tennessee. She has a diverse background in marketing,

purchasing and accounting.



Marilyn lives in Camden, Arkansas and is serving Alabama, Arkansas, Louisiana, Oklahoma and Mississippi and has professional experience in customer service and marketing.

AlphaNet has trained both Jane and Marilyn in our health management program, our support systems and infusion supply ordering procedures. We extend a warm welcome to both as they use their enthusiasm, skills, experience and insight as Alphas in service to the Alpha-1 community.



Flying High

Mary Pierce, AlphaNet Coordinator, has a new passion – flying a powered paraglider. Mary is pictured here during her first solo flight. She says when her flying skills improve she wants to paint “Cure A-1” on her wing.

Pulmonary Hypertension in Alpha-1

By Robert A. Sandhaus, MD, PhD, FCCP - Medical Director, AlphaNet

Note: This is not an exhaustive discussion of pulmonary hypertension in general but limited to the major issues related to this condition faced by the Alpha-1 community.

Nearly everyone has heard the fancy medical word for high blood pressure: *hypertension*. This very common condition is easy to diagnose by wrapping a blood pressure cuff around someone's arm and taking a reading. But many Alphas with lung disease suffer from a much more difficult to diagnose type of hypertension that can have serious health consequences if not identified: *pulmonary hypertension*.

Pulmonary hypertension is an elevation of the pressures in the blood vessels feeding the lungs. This often occurs without any elevation in the blood pressure in the rest of the body. When significant pulmonary hypertension is present it can lead to a variety of symptoms including increased shortness of breath often with exertion, chest pain, liver swelling, fluid accumulation in the legs or abdomen, and a form of heart failure known as cor pulmonale (see below). A major problem with diagnosis is that when pulmonary hypertension is still in its early stages and the pressures in the blood vessels are only mildly elevated, there are often no symptoms at all. The earlier that pulmonary hypertension is identified, the easier it is to treat.

So what is the relationship between pulmonary hypertension and Alpha-1?

Alpha-1 itself does not directly lead to pulmonary hypertension but individuals with lung disease due to Alpha-1 are at risk of a form of this condition known as secondary pulmonary hypertension. Secondary pulmonary hypertension is due to prolonged low oxygen levels in the lung. The lung has a built-in mechanism that is designed to close down the blood supply

to parts of the lung that are not receiving enough oxygen. Presumably, this is a defense mechanism that would shunt blood away from a section of the lungs that might have a blocked airway (caused by aspiration of a foreign body like a peanut, for example) or a damaged area of lung (such as in a severe pneumonia localized to one or two lobes of the lung). Closing down these blood vessels will effectively shunt the blood supply of lung away from these ineffective areas to more normal areas of the lung, allowing the blood to continue to pick up normal amounts of oxygen.

But what happens if the damage to the lungs and low oxygen levels are spread throughout the lung as in someone with the generalized emphysema of Alpha-1?

In that case, all the blood vessels in the lung start to spasm and close down. This results in elevation of the pressures of the major blood vessels that bring blood into the lungs from the right side of the heart, leading to pulmonary hypertension. It is called secondary pulmonary hypertension in this setting because the elevated pressures are the result of (or secondary to) the low oxygen levels in the lungs.

When pulmonary hypertension becomes severe, it strains the function of the right side of the heart. The right heart chambers enlarge or dilate, and if this continues long enough, the pumping action of that side of the heart becomes much less effective. This is called 'right heart failure.' When right heart failure is due to pulmonary hypertension from lung disease this is known as cor pulmonale (pronounced core'-pul-mon-a'-ly). When the right side of the heart fails, fluid "backs-up" into the veins that feed the right heart. These veins normally drain the extremities and the abdomen so the result can be fluid (or edema) swelling the liver, the ankles and legs, and very full jugular veins in the neck.

CLINICAL CORNER

Considerations for Managing Alpha-1 Augmentation Therapy At Home

By Kathy McKay, RN
AlphaNet Clinical Nurse Manager

If you receive augmentation therapy at home, here is some information you should know:

- Be sure to unpack your box of medication as soon as it arrives and reconcile its contents with the packing slip.
- Inspect the shipment for any damage and call your AlphaNet Coordinator immediately if the contents are in any way compromised.

The good news about this type of secondary pulmonary hypertension is that maintaining adequate oxygen levels with supplemental oxygen can prevent it from ever occurring. Individuals with early signs of pulmonary hypertension can reverse these changes by starting supplemental oxygen in a timely fashion. Even those with advanced cor pulmonale will benefit from returning their oxygen levels back toward normal by using supplemental oxygen 24 hours a day.

So how do you know if you have pulmonary hypertension?

There is no simple office test that can make the diagnosis. If someone with lung disease and low oxygen begins to show signs of right heart failure, then pulmonary hypertension is likely. But it is important to make this diagnosis before such heart effects are seen.

There are some changes that can be seen in a simple EKG when pulmonary hypertension strains the heart. The most common non-invasive method to evaluate the blood pressure in the pulmonary arteries is a cardiac echo or ultrasound, also called an echocardiogram. The echocardiogram can look for enlargement of the pulmonary artery and can also make indirect measurements of pulmonary artery pressure. The problem with the echocardiogram is that air in the lungs can block the sound waves that produce the image of an echocardiogram. People

with lung disease due to the Alpha-1 usually have hyperexpanded lungs and air trapping that make imaging the pulmonary arteries difficult.

Another indirect method of looking for pulmonary hypertension is a chest x-ray or CAT scan of the chest. These types of x-ray studies produce images of the pulmonary arteries that can be evaluated for enlargement of these vessels which can be an early sign of increased blood pressure.

Finally, the “gold standard” for evaluating pulmonary artery blood pressure is a heart catheterization. This technique involves threading a catheter into one of the main veins draining the leg, arm or neck and advancing it into the heart to measure pressure directly. This is more invasive and is often not needed in the setting of secondary pulmonary hypertension, but may be required if other techniques don't give a reliable answer.

If there is a bottom line to this story, it is a reinforcement that when supplemental oxygen is indicated for

patients with lung disease due to Alpha-1 because of low oxygen levels at rest, with exercise, or during sleep, its use will not only improve your ability to function today but it will also prevent the more serious complication of secondary pulmonary hypertension in the future.

*There is also a form of pulmonary hypertension known as **primary pulmonary hypertension**. This is often a serious medical condition in which there are abnormalities of the small blood vessels within the lungs leading to elevated pressures in the pulmonary arteries without any apparent lung disease. This can be life threatening but, fortunately, there are a variety of new therapies that are available for this population. There is no evidence that people with Alpha-1 are at higher risk of developing this form of pulmonary hypertension.*

- Store the medication according to the labeled instructions. Alpha-1 augmentation therapy drugs can be stored at temperatures not to exceed 77° F. (25° C). These drugs are safely stored at room temperature, but you'll need to check the information provided on your specific drug package insert for the appropriate length on time. If you have any questions or concerns, please consult your pharmacist.
- Your medication should never be frozen. The vial of diluent (sterile water) might break.
- If you refrigerate your medication-be sure to remove it from the refrigerator in plenty of time for it to warm to room temperature prior to your infusion. One suggestion is to remove it the night before.
- Never refrigerate your medication after it has been prepared or “reconstituted.”
- It is important to read the prescription label with each delivery you receive, since the number of vials that will be prepared to achieve your prescribed dose CAN CHANGE. Read the labels carefully and be sure your infusion nurse or caregiver checks the labels each time when preparing your infusion.
- Remember that your infusion must take place within three hours after the medication is reconstituted, as it contains no preservatives. To avoid any problems related to this time constraint, it is recommended your nurse establishes your IV access prior to preparing the drug. This prevents having to play “beat the clock” if any problems occur in starting your IV.

If you have a clinical topic you think all Alphas should learn about in the next issue of AlphaNetter, please contact Kathy McKay at kmckay@alphanet.org or 877-913-8141.

CAREGIVERS ALSO NEED SPECIAL CARE

AlphaNet Coordinator Kathy Haduck has first-hand experience of just how physically and mentally debilitating it is to be a caregiver to someone critically ill. The isolation, fear and self-sacrifice of caring for her dying sister resulted in Kathy ignoring her own serious health emergency putting her in the trauma unit within minutes of her sister's death.

"Every second of every day was consumed with my sister's needs," Kathy explained. "And though those seconds, minutes, hours, days, and weeks were freely and lovingly given, I look back amazed that I survived it all. I certainly didn't take the time I needed to recharge my batteries and emotions."

Kathy has come across "A Caregivers Bill of Rights" and suggests everyone keep a copy and share it with their loved ones. She has placed hers in her Bible for safekeeping.

A Caregiver's Bill of Rights

I have the right...

- ...to take care of myself. This is not an act of selfishness. It will give me the capability of taking better care of my relative/loved one.
- ...to seek help from others even though my relatives may object. I recognize the limits of my own endurance and strength.
- ...to maintain facets of my own life that do not include the person I care for; just as I would if he or she were healthy.
- ...to get angry, be depressed, and express other difficult feelings occasionally.
- ...to reject any attempt by my relative either conscious or unconscious to manipulate me through guilt, anger or depression.
- ...to receive consideration, affection, forgiveness and acceptance for what I do for my loved one for as long as I offer these qualities in return.
- ...to take pride in what I am accomplishing and to applaud the courage it has sometimes taken to meet the needs of my relative.
- ...to protect my individuality and my right to make a life for myself that will sustain me in the time when my relative no longer needs my help.

An adaptation of the Family Pledge of Nonviolence created by the Institute for Peace and Justice (www.ipj-pj.org) and reprinted with their permission.



Reeling Them In

Chuck Cook, an Alpha from Oak Lawn, Illinois, is Chairman of the Fish Tales of Oak Forest fishing club with 150 members including AlphaNet Coordinator, Liz Veronda. He leads regular trips to fish the waters around his area and to far flung destinations such as Canada, Hawaii and the Dry Tortugas (off the Florida Keys). He caught this walleye on a recent trip, with seven club members, to Little Bay de Nock in the upper peninsula of Michigan, in 14-degree weather. A retired mechanic for UPS, Chuck was diagnosed with Alpha-1 ten years ago.

Enterprising Alpha Finds Winning Solutions

Since her diagnosis with Alpha-1 seven years ago at age 37, Julie Brittingham has noticed a hyper-sensitivity to environmental factors, including cleaning products. With just 22% lung capacity at diagnosis, Julie struggled for years to determine the causes of her varied allergic reactions. Even during hospitalization, she would react to unknown factors. Eventually, by using herself as a test subject, she narrowed the problem to things around her in her home environment.



Julie surrounded by her new line of hypo-allergenic cleaning products.

"It is so important to eliminate as many toxic triggers as we can in our home and workplace because irritation to our lungs can result in potential damage," said Julie.

She kept researching and trying different products. Finally, too debilitated to clean her home herself, she hired a cleaning service and things got even worse. She had to leave while they cleaned. Four years ago she found one all-purpose cleaning product through an allergy magazine. The product was a winner: she didn't have a bad reaction. Still, it took several more years of reacting to other products around her home, and dead ends with her research, before Julie took charge.

After Julie went out to eat Chinese food with a friend and got a fortune cookie that said, "be innovative and take charge of new ideas", she finally did take charge. She decided she needed to come up with a plan to solve her problems, help others, and make enough money to move out of her old home into one that would have fewer environmental issues.

Julie went home and looked on the back of the bottle of the only product that had ever worked for her. She found the manufacturer and after a few weeks of persistent phone calls, she tracked down who was behind the product. This cleaning solution had been developed by a chemical engineer, with extreme chemical sensitivities, who had designed it for himself. Coincidentally, he was just planning to launch an entire new line of cleaning products for both home and commercial use. Julie moved quickly to become involved and has set up a company, JULSHSCS, to market the non-toxic cleaning product line Alpha Health Smart Cleaners.

To contact Julie, call toll-free 866-448-A1AD (2123) or look for her products on ebay by searching for "Alpha Health Smart Cleaners." She plans to donate 10% of her proceeds to the Alpha-1 Foundation.

LOCATED AT THE UNIVERSITY OF FLORIDA

Alpha-1 Foundation DNA and Tissue Bank

The Alpha-1 Foundation developed the DNA and Tissue Bank, in part with funds donated by AlphaNet, to provide a repository of DNA and tissue samples for Alpha-1 research. The Tissue Bank Advisory Committee, a group composed of researchers and Alphas, operates the bank on behalf of the Alpha-1 Foundation. Researchers investigating Alpha-1 Antitrypsin Deficiency (Alpha-1) and other disorders, use the Bank to further scientific and medical research.

Participation in the DNA and Tissue Bank is optional and open to both Alphas and non-Alphas. Participants grant informed consent, participate in a survey, and then are given detailed instructions on providing either a blood sample or tissue obtained during biopsy or transplant. People who have previously donated a blood sample or tissue sample may contact the DNA and Tissue Bank to update valuable medical information, such as pulmonary function test results.

The DNA and Tissue Bank is operated under stringent federal laws and is overseen by an Institutional Review Board (IRB). Personal health information and records are used only by Bank staff. No private data or records such as name, address or social security number are ever given to any researcher. Only researchers whose projects have been approved by the Tissue Bank Advisory Committee have access to coded samples, tissue and/or medical information.

Genetic data may advance research into more effective treatments and potentially, a cure. Alphas who donate blood and tissue are forging a partnership with the Alpha-1 Foundation and with scientists researching Alpha-1.

FOR MORE INFORMATION, PLEASE CONTACT:

Alpha-1 Foundation's DNA and Tissue Bank
University of Florida
Mark L. Brantly, MD
Principal Investigator
Toll Free Phone: 1-866-284-2708 (option 3)
Email: alpha1lab@alphaone.ufl.edu

www.alphaone.ufl.edu

Avid hunter, fisherman and AlphaNet Coordinator, Liz Veronda, poses after a successful winter hunt with Jeff Foiles of Foiles Migrators, a company that makes world champion duck and goose calls.



Into The Wild

Indiana, PA Support Group Meeting with Kathy Haduck, AlphaNet Coordinator; Jim Cantwell, CHR VP Marketing; Dana Myers, Talecris Rep; Ashok Chainani, Talecris Director of Managed Care and Reimbursement Strategy; Cathy Gould, Support Group Leader and Dottie Condon, group member and guest speaker for the event which drew nearly 30 participants.



Indiana PA Support Group

**COME SEE OUR WEBSITE'S
NEW LOOK**

If you haven't visited the AlphaNet website in a while you are in for a surprise. The site has been revamped and updated with more dynamic information, news updates and easy to access information. Stop by today:
www.alphanet.org

April 7

11th Annual Clean Air Bike Ride / American Lung Association.

**Withlacoochee State Trail through Central Florida
Registration begins January 1, 2007 for 12, 20, 48
or 100 mile rides.**

**Contact AlphaNet
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866-762-1850**



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