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# The AlphaNetter



## Alpha Breeder Keeps Ducks in a Row

Professional breeder and Alpha, Jane Edington, knows how to keep her ducks in a row. She has been raising British waterfowl, breeding schnauzers and grooming dogs for nearly 50 years. Jane resides in Hollis Center, Maine and is a member of the British Call Duck Club and District Director of the National Call Breeders of America. In 2008, she was a call duck judge for the Welsh National in Great Britain. When prompted to expound on her accomplishments, Jane is quick to respond, "I live with a lot of ducks, some pups, and a team of horses. The kids are with me now too. We're just putting one foot in front of the other and taking it a day at a time."

"Taking-it-a-day-at-a-time" involves waking up at 5:30 a.m. to water 70 breeders and the 147 ducklings Jane hatched this spring, looking after two new schnauzer pups, and delivering ducks as far as Boston, New Hampshire, New York and Europe. Jane

squeezes in time between mending the fence and feeding the horses to explain the finer points of imprinting, taking a duck away from its natural parent so that you become the parent, and the standard differences between the European Peking duck and the U.S. variety.

Jane's mother was a Harvard graduate with a passion for genetics and color breeding daushaunds. When Jane went to live with her at age 19, she caught the breeding bug and joined the business. In 1960 the mother-daughter pair went to pay their condolences at a nearby kennel that had caught fire. There Jane bought her first five schnauzer pups using part of her inheritance from her grandmother. She has bred schnauzers ever since.

Jane's interest in waterfowl developed later. In 1967 she bought her first pair of spot call ducks. Call ducks were a popular hunting decoy in Holland in the 1600s and continue to be used for hunting, show and as pets.

"It took awhile to learn all the science and standards related to breeding, but eventually it clicked. It's a great hobby and a fun community to be involved with," Jane says.

Jane was diagnosed with Alpha-1 in 2002 and began augmentation therapy. "I never could blow up balloons as a kid," Jane quips, "but honestly, I don't get too caught up in the disease. You have to keep going and I have plenty to keep me going. The truth is your health is never trustworthy. I'm a person of faith. That's what I trust in and faith and family have made my life full."



Jane's Schnauzer pups and baby ducklings





# Oxygen

## The Gas We Love to Hate

By Dr. Robert A. Sandhaus, MD, PhD, FCCP

Oxygen is a wonderful gas. It's all around us. It's in every breath we take. Not only that, we're all addicted to oxygen. If oxygen were to be reduced or removed from the air, we would all experience extreme, even fatal, withdrawal symptoms!

One of the primary jobs of the lungs is to take the oxygen we breathe and deliver it to the blood. Oxygen-rich blood can then bathe all the tissue of the body and essentially feed these tissues. Oxygen, one of our primary nutrients, allows the organs of our body to function and use fuels such as sugars, fats and proteins. Without oxygen, our muscles (including the heart), brain, liver and intestines cannot function for long. When these organs use oxygen and fuel, they generate the primary waste product carbon dioxide. The lungs have the job of removing carbon dioxide from the blood; we then exhale it as we breathe.

For some people with lung disease, the damage to the lungs is severe enough to reduce the amount of oxygen delivered to the blood. Reduced oxygen can cause various tissues of the body to become starved for oxygen and function poorly. The blood vessels in the lungs narrow and make it difficult for blood to flow through. This may put strain on the right side of the heart, which delivers blood to the lungs.

The most direct therapy to improve oxygen levels in people with lung disease is to treat the underlying lung disease. But while the underlying lung disease, such as pneumonia, is being treated, it may be necessary to temporarily give that person a higher concentration of oxygen to breathe. Supplemental oxygen helps to make up for the lower delivery of oxygen by the injured lung tissue and can be delivered by nasal cannula, oxygen mask, oxygen tent (not used anymore), or via a tube inserted into the windpipe if their pneumonia is extremely severe.

Individuals with low oxygen levels due to lung disease that can't be cured or reversed may need to use oxygen on a regular or even continuous basis. This is often the case with people who have severe emphysema due to alpha-1 antitrypsin deficiency (Alpha-1). Even with maximal medical therapy, some Alphas have such severe lung disease that their low oxygen levels put the other organs of the body at risk of injury. Low oxygen levels may occur only under certain circumstances, such as when they sleep, exercise, eat, talk, are up and about, or even while simply sitting doing nothing. In general, people with Alpha-1 and low oxygen levels only need to use oxygen at times when they know their oxygen levels are likely to be low.

Oxygen levels can be checked using an oximeter or by doing an arterial blood gas (ABG). An oximeter shines a light of a specific color through the skin and uses a sensor, that is placed on the finger or ear lobe, to measure the changes in the amount of light that is absorbed as the blood pulses through the underlying blood vessels. The readout of an oximeter is a percentage from zero to 100 that represents the percent of hemoglobin molecules in the red blood cells that are carrying oxygen. In normal people at sea level, close to 100% of the hemoglobin molecules in the blood will be carrying oxygen – at higher altitudes this number drops because there is less oxygen available in the air as you get higher and higher in the atmosphere. Currently, physicians consider oximetry readings of less than 88% to be too low for good health.

An ABG is measured by drawing blood from an artery, usually in the wrist, and analyzing it with a machine that measures oxygen, carbon dioxide, the acid-base balance of the blood (pH) and several other parameters. While this is more "invasive" than a simple sensor worn on the finger, it provides considerable additional information. A physician may initially order an ABG and follow the patient in the future with oximetry alone.

(Continued on adjoining page)

(OXYGEN Continued)

Suppose your oxygen level drops below an oximetry value of 88% when you sleep, exercise, or all the time. After your healthcare provider has done everything that can be done to improve your lung function, if the oxygen level is still too low, they will likely recommend supplemental oxygen. Most people receive their oxygen by a nasal cannula attached to an oxygen source. This source can be an oxygen tank with compressed oxygen gas in it, a liquid oxygen tank or portable unit, or an oxygen concentrator, which pulls oxygen directly from the air around it. Instead of a nasal cannula (sometimes called nasal prongs), some patients use an oxygen mask, "oxygen eyeglasses" (in which small nasal prongs protrude from the frame of eyeglasses), or transtracheal oxygen (in which a small tube is inserted surgically directly into the windpipe and this tube is used to deliver oxygen). There are benefits and disadvantages to each of the different oxygen delivery systems.

People that have to exercise or walk with their oxygen generally use a nasal cannula attached to a compressed

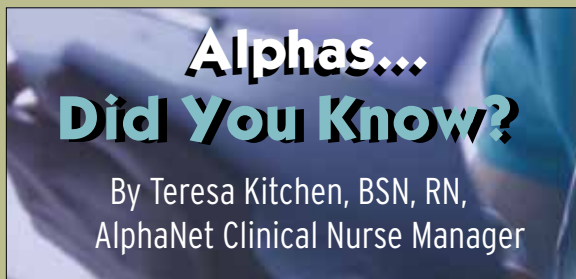
gas oxygen tank (there are a variety of sizes of these) or a portable liquid oxygen system. The standard way of determining the amount of oxygen delivered is by the number of liters of oxygen gas that flows through the cannula in a minute (L/min) as a continuous flow. Another method of sending oxygen through the tubing to the nose is the pulsed delivery system. Pulsed delivery only sends oxygen through the tubing when the unit senses that you are inhaling and turns off the oxygen during the time you are exhaling. This method is intended to allow a user to go for a longer period of time between refills or tank changes. Not all patients can maintain their oxygen levels on pulsed delivery systems.

Often people are concerned that if they breathe through their mouth or have a stuffed nostril, when using a nasal cannula, they won't get the oxygen they need. Basically, the nasal cannula uses your nose and the back of your throat as a reservoir to hold oxygen when you're not inhaling, so even mouth-breathers can improve their oxygen levels using a nasal cannula. If one of your nostrils is congested or closed, you may have to turn up your

oxygen a bit, but usually not much. On the other hand, there are certain pulsed delivery systems that use one nostril to deliver oxygen and the other to sense when someone is inhaling (so a pulse of oxygen can be delivered). A clogged nostril can lead to a significant drop in oxygen delivery in that setting.

If your doctor tells you that you need to wear oxygen, either 24 hours a day or in certain situations, your first reaction, if you're like most people, is to panic. What will my spouse think? What will my boss, coworkers, and clients think? Everyone will stare at me. Everyone will know I'm sick. I'll become addicted to this oxygen and will never be able to get it off.

This last is easy to answer. As you know, we're all addicted to oxygen. None of us will ever "get off" oxygen until death. You just need a little bit more oxygen than those with normal lungs. The other questions you ask yourself don't have simple answers, except to say that all of those concerns won't matter to you if you wind up in the hospital in heart failure because your low oxygen level caused the other organs of your body to fail.



- The Alpha-1 Association 20th Annual National Conference is set for June 10-12, 2011 in St. Paul, Minnesota.
- To obtain a free coded fingerstick test kit for Alpha-1 contact the Registry Coordinator toll free at 877-886-2383.
- It is strongly recommended that all individuals diagnosed with COPD be tested for Alpha-1.

- When you have any change in your insurance coverage you should notify your physician's office, hospital, pharmacy and home care company PRIOR to the effective date of the change.
- Research your insurance options closely with COBRA or Medicare. Seek out assistance and know your rights.
- Attempt to empty the IV tubing at the end of your infusion to obtain the most of your augmentation therapy.
- Portable oxygen units should be checked periodically by the oxygen provider to make sure the unit is working properly.
- It is recommended that FluMist NOT be used by Alphas with lung disease or by family members and others who have close contact with Alphas with lung disease; FluMist is a LIVE virus.



# Navigate Alpha-1 with Our Unparalleled Health Management



*AlphaNet Coordinators, Board members and staff gathered in Orlando in June to attend the Alpha-1 Association's National Education Conference and to receive additional training.*

## **Know Your Resources**

AlphaNet is pleased to offer personalized health management services to Alphas living in the United States. All enrolled AlphaNet subscribers are part of our Alpha-1 Disease Management and Prevention Program (ADMAP). This comprehensive plan, designed to assess and monitor patient health and prevent disease progression, is part of AlphaNet's standard of care for all subscribers and includes the following components:

### **The Big Fat Reference Guide (BFRG)**

The *BFRG* is truly the most comprehensive and accessible publication for Alphas. Its content spans basic information on Alpha-1, health management for lung and liver disease, organ transplantation and more. The *BFRG* is based on published scientific evidence and sound clinical practice. It is available, along with the *Skinny Little Reference Guide (SLRG)* collection, to all Alphas online at [www.alphanet.org](http://www.alphanet.org)

### **Monitoring by AlphaNet Patient Service Coordinators**

All AlphaNet subscribers are assigned a Coordinator based on geographical location. Coordinators are Alphas who have been trained in AlphaNet's health management program. Your Coordinator will contact you on a monthly basis to offer support and education, gather data, keep you informed about current research studies, and guide you through collaborative self management. Our monthly questionnaires will help you track your progress and identify areas to improve. Your participation is crucial to your individual success in the program. Be sure to return missed calls from your Coordinator and other AlphaNet staff at your earliest possible convenience.

### **Outcomes**

The effectiveness of the ADMAP program was evaluated during an outcome study conducted from January 2003 through July 2005. The results indicated that Alphas with lung disease, who are followed by AlphaNet, maintain their quality of life and experience fewer, shorter and less severe exacerbations. In addition, they use less antibiotics, have fewer visits to the emergency room and spend less time in the hospital. Our latest effort to improve our health management services, to identify interventions to incorporate into our program and to acquire important data about Alpha-1, is the Step Forward Study (SFS), a 5-year study that began in 2009.

### **Education**

AlphaNet provides professional education opportunities to RNs, physicians and respiratory therapists online. Programs are approved for three continuing nursing education credits and three continuing medical education credits.

Visit [www.alphanet.org](http://www.alphanet.org)

## Treatment Planning and Review

AlphaNet's medical team provides treatment planning and review services to AlphaNet subscribers. This process involves asking the participating physicians to send a copy of the patient's treatment plan to AlphaNet for review. The medical team at AlphaNet then works with the physician to ensure that the treatment plan meets the Standard of Care for Alpha-1. Support and resources are made available to the many physicians whose clinical practice includes limited experience with Alpha-1. Emphasis is on securing the physician's participation, encouraging adherence to guidelines and decreasing the significant variations in care for the Alpha-1 patient.

## In Memory of Marta Strock December 8, 1949 - June 2, 2010



Marta Strock was one of the original AlphaNet Coordinators and a beloved member of the AlphaNet community. She served from 1999 until 2009, first as Coordinator for Florida and later as At-Large Coordinator. In addition to serving at AlphaNet, Marta helped to set up the Miami offices for the Alpha-1 Foundation and served on the Board of the Alpha-1 Association Florida Chapter.

In 2001, Marta underwent surgery for breast cancer, and she retired from AlphaNet in 2009 due to illness. Marta passed away peacefully on June 2, 2010. She is survived by her life partner Sue Fair and her mother, Claudel Strock.

## New Program Offers CRC Access

In May, Alpha Molly Hunter traveled to National Jewish Health Medical Center in Denver, Colorado for evaluation and testing as a part of the Alpha-1 Foundation's Clinical Resource Center Access Program (CRC). Molly learned about the opportunity while discussing an ongoing battle with the bacteria Mycobacterium Avium Complex (MAC), in addition to her Alpha-1, with her AlphaNet Coordinator, Barb Pusey. Molly was the first Alpha to take advantage of this program.

"My doctor told me that National Jewish was the most knowledgeable respiratory hospital and that it was top-ranked in the nation. Barb explained how the CRC Access program could help me be evaluated there. I applied, was granted a travel stipend, and was not disappointed," Molly says.

The medical team at National Jewish was able to diagnose several other conditions that were affecting Molly's breathing and had until then gone undetected. Molly reports that she has followed their recommendations and is breathing much better since returning home.

"In addition to being prescribed a new inhaler, I also learned that, by failing to clean the tubing of the nebulizer that I previously used three times a day, I may have been re-infecting myself. I use it less now, but I am sure to use a new or clean tube each time. All the little things that I have been doing since I came back have helped me tremendously. I encourage other interested Alphas to talk with their Coordinator. Getting the right diagnosis is the most important thing you can do - than you can get the right treatment," Molly says.

The Alpha-1 Foundation provides travel stipends to assist newly-diagnosed Alphas and their caregiver to visit their closest CRC. Alphas have the opportunity to be evaluated by a physician who is familiar with Alpha-1, ask questions and receive answers regarding their condition. Alphas must be recommended by their AlphaNet Coordinator and this must be their first time visiting a CRC. Stipends are based on distance per roundtrip travel.

There are over 60 CRC's throughout North America that specialize in patient care and education for those with Alpha-1. Some centers treat lung disease and others liver disease. Centers have other resources for Alphas such as support groups, transplant centers and pulmonary rehabilitation.



## Alpha-1 Association Education Days

co-sponsored by  
the Alpha-1 Foundation

October 23  
Seattle, Washington

November 6  
Temple, Texas

December 4  
Las Vegas, Nevada

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

Your AlphaNet Coordinator is a great resource, whether you are a newly-diagnosed or long-time patient. Take advantage of his or her expertise. Check out "Questions to Ask Your Coordinator" and other FAQs at [www.alphanet.org](http://www.alphanet.org)

## Alpha-1 Foundation Event Calendar

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



October 9 - Brookside Croquet  
Championship  
In Honor of Todd Zinni  
S. Nyack, New York

November 20 - Alpha-15k Walk Miami  
Miami, Florida

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

### Alpha-1 Business Updates

In June 2010, Grifols, SA, a Spanish pharmaceutical company, announced its intention to acquire Talecris Biotherapeutics, a global biotherapeutic and biotechnology company and manufacturer of ProLasta<sup>®</sup>.

In August 2010, Baxter International entered into an agreement with Kamada, Ltd. to become the sole distributor of Glassia<sup>™</sup>, a liquid alpha-1-proteinase inhibitor produced by Kamada. Glassia<sup>™</sup> was approved by the FDA on July 1, 2010.



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