ALPHA-1 FOUNDATION RESEARCH REGISTRY UPDATE

MUSC MEDICAL UNIVERSITY of SOUTH CAROLINA

Vol. 11 / Issue 1 / Winter/Spring 2009

Letter from the Director

By Charlie Strange, MD
Professor of Pulmonary, Critical Care, Allergy and Sleep Medicine
Medical University of South Carolina

Dear Registry Members,

WELCOME to this edition of the Alpha-1 Foundation Research Registry Newsletter. Inside you will become acquainted with three recently-added clinical resource centers, all in Chicago.

Does the Alpha-1 community really need three centers in Chicago? My answer is, absolutely! First, Chicago is a big place. More importantly, the more physicians who know about Alpha-1 Antitrypsin Deficiency and have an interest in caring for patients with Alpha-1, the better.

One of the new initiatives of the Alpha-1 Foundation is to find knowledgeable physicians accessible to all individuals with this condition. The Research Registry has collected the names of your Alpha-1 physicians for the past 12 years. We have never reached out to them, since the majority of physicians listed in the Registry care for a single family with this condition and the confidentiality of the Registry participants has always been the Registry's foremost concern.

However, the time has come to improve community care for Alphas, and new initiatives are in the making for expanding the network of physicians that know this condition well. If you think your physician does a great job caring for Alpha-1 patients we want to hear about it. Please contact the Alpha-1 Foundation at rasandhaus@alphaone.org; the Foundation will reach out to them and ask if they have an interest in becoming an Alpha-1 Clinical Resource Center.

The Registry Staff continue to streamline their procedures for testing in the Alpha Coded Testing Study. We think you will find our featured story in this issue about Alpha-1 testing after liver transplantation an interesting way to improve your personal knowledge about testing. The Registry staff, Rebecca McClure and Laura Schwarz, and Dawn McGee, the genetic counselor for the Alpha-1 Association, are available to discuss issues about testing from 8-5 Monday through Friday at our toll-free numbers. In addition, more reading on testing is available in The Big Fat Reference Guide online. For those who have not yet found it, please visit www.alphanet.org.

In this newsletter, those of you who have told us that you are genotype PiZZ will find a study invitation from Friedrich Kueppers, MD, Professor of Medicine and Microbiology/Immunology at Temple University in Philadelphia. Sometimes the best studies are the most simple. Dr. Keuppers would like to know the number of children born to your grandparents and compare this number to children of the grandparents of your current or last spouse. His invitation letter describes the research question in more detail. We expect his limited questions will generate some clarifying discussions that we encourage you to ask of the Registry staff.

Some of you may be aware that the Research Registry has an oversight committee, which assures that research requests have met necessary standards for research. This committee has been chaired for the past two years by David Coultas, MD, Chairman of the Department of Internal Medicine at the University of Texas Health Sciences Center at Tyler. As he steps down from this role, we should all thank him for the job he has done. We welcome Jim Stocks, MD, Professor of Pulmonary and Critical Care Medicine in Tyler, TX, back to this important job as the new chair of the Research Registry Oversight Committee.

Please note: some new research studies are close to beginning enrollment. Please consider these studies when the invitation arrives in your mail.

Sincerely,

Charlie Strange

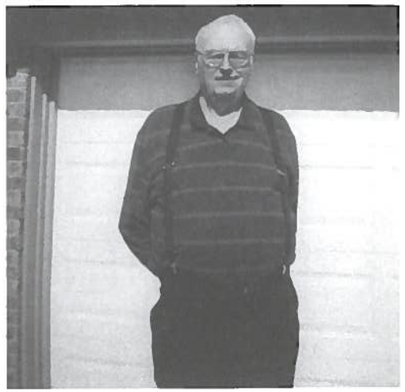
Charlie Strange, MD
Director, Alpha-1 Foundation
Research Registry

Is he MM or ZZ? Or is he just from Texas?

By Laura Schwarz (with technical assistance from Charlie Strange, MD)

LAST SPRING I received a call from a man in Texas named Jim Franklin. He had received a letter from the Registry advising him that our blood test showed his genotype as ZZ. He wanted to know how he could be a ZZ, since six months before receiving our letter, he had a phenotype performed at Baylor All Saints Medical Center in Fort Worth – and the result showed his phenotype is MM.

I was perplexed and became even more so when I saw that his protein level was in the normal range at 35 μM! Something wasn't right.



Jim Franklin

Here is some background on Jim Franklin.

Franklin was born in Cleveland, Ohio, in 1942. He joined the Air Force in 1961 and remained in the service for 22 years. He was based all over the country and elsewhere. In 1978, he was stationed at Carswell AFB in Fort Worth. He met his wife, Carol, in San Antonio and they married a year later. The Franklins, along with his two sons and her son and two daughters, established their home in Fort Worth. In 1983, he retired from the Air Force and became a technical writer for General Dynamics, which later became Lockheed Martin, writing avionics/weapons system repair manuals and flight/weapons delivery manuals for the F-16 Fighting Falcon. He retired in 2007.

There are three common tests used to detect alpha-1 antitrypsin deficiency (Alpha-1).

Franklin had had no serious health problems until the age of 47, when he was admitted to the hospital with a fever and back pain and was found to have gallstones. Upon removing his gallbladder, the surgeon found cirrhosis in his liver. Franklin was stunned; he had never been a heavy drinker. He persistently returned to the doctor's office, hoping to find the cause of his liver disease.

Two years later, in 1991, he had a liver biopsy at All Saints Hospital. He was diagnosed with cirrhosis related to Alpha-1 Antitrypsin Deficiency. He was told very little else, as no one seemed to know much about the condition.

He changed doctors, and his new physician surprised him by asking if he had considered a liver transplant. He received a successful liver transplant at Baylor Medical Center in Dallas Sept. 8, 1993. The next 14 years went well except for double hip replacements in 2004-2005.

In 2007, Franklin decided to get tested again for Alpha-1. He wanted to set a good example for his children, and either did not remember or had never been told the specific results from his Alpha-1 tests from 1991. He asked his pulmonary specialist to retest him. The result showed an MM (normal) phenotype!

This result confused him, and when, six months later, he heard about the Alpha Coded Testing (ACT) Study through his sister, he requested a fingerstick test.

That was the test that led to our letter identifying him as Alpha-1 deficient with a ZZ genotype – and an estimated protein level of 35 μM, which is in the normal range. Quite reasonably, Franklin called the Registry to question the two different test results.

We tell his story to educate everyone about this unusual situation.

There are three common tests used to detect Alpha-1 Antitrypsin Deficiency (Alpha-1).

The most commonly used test is a test of the amount of alpha-1 antitrypsin in the blood.

The most commonly used is a test of the amount of alpha-1 protein in the blood. This is usually called an Alpha-1 concentration or alpha-1 level. This test result is given in either micromolar (μM) or milligram per deciliter (mg/dl) levels. Levels less than 11 μM or 52 mg/dl are used to identify patients at highest risk for lung and liver disease.

The second test is called a phenotype.

The second test is called a **phenotype**.This test collects blood and isolates the alpha-1 protein from all the other blood proteins. This protein is then placed on a gel (it looks like a plate of clear Jell-O) and an electric current is applied that isolates the protein bands into the different types. M protein, S protein, and Z protein all move to different places on the gel, and when stained, can show the type of Alpha-1 protein being produced by the liver.

The third test is called a genotype.

The third test is called a **genotype**. This test uses white blood cells, which have a nucleus (red blood cells do not). The nucleus is where our chromosomes live. These chromosomes carry all of our genes. The gene for Alpha-1 can be searched for by a very sensitive test that finds all of the S and Z genes and determines if one or two copies of these genes are present. Since everyone has two copies of the Alpha-1 gene called SERPINA1, two letters for this gene are reported. Importantly, the genotype is testing blood cells that come from the bone marrow – ***not*** from the liver – so the test is not affected by a transplanted liver. Because the genotype test requires only small amounts of blood, it can be performed by fingerstick testing.

Our genes don't change anywhere in the body except for a transplanted organ.

With this information, Franklin's test results are easy to explain. His liver (from his liver donor) had normal MM cells and was making MM Alpha-1 protein in normal amounts. This gave him a normal blood level of Alpha-1 (35 μM) and a phenotype of MM protein. However, a gene-based test from anywhere else in his body would show that everywhere except the liver, he would have the same genes that were present when he was born. In this case the blood genotype showed Franklin to be ZZ. Our genes don't change anywhere in the body, except for the transplanted organ.

Last year, Franklin began having breathing problems when he was in the hospital for surgery. His current pulmonologist is treating him for chronic obstructive pulmonary disease (COPD) and for obstructive sleep apnea syndrome. Franklin takes medications, oxygen at night, and uses a mask to ventilate with a bi-level positive airway pressure (BIPAP) machine. Although his COPD is attributed to Alpha-1, Franklin will not benefit from augmentation therapy, since his blood levels of Alpha-1 are already at the normal level because of his 1991 liver transplant.

The Alpha-1 Registry wishes Jim Franklin well – and suggests that other liver transplant recipients not repeat this long and confusing course!

Winter/Spring 2009 Research Registry Update Contents

You may contact the Alpha-1 Foundation Research Registry staff by email, at alphaone@musc.edu for additional assistance in locating resources related to AAT Deficiency research, to obtain information about current research activities, to participate in the Research Network or Registry, or to receive Foundation publications.

News from Capitol Hill

New Congress likely to tackle healthcare reform in 2009

By Miriam O'Day
Senior Director of Public Policy, Alpha-1 Foundation

MANY PROBLEMS have consumed the presidential election cycle and the attention of Congress at the end of 2008. In spite of this crisis-solving agenda, healthcare and the opportunity to solve some of the current healthcare problems is at the top of the list of things to be addressed in the beginning of the 111th Congress, which convenes in January 2009.

There is much excitement about the opportunity for comprehensive healthcare reform in Washington, as many believe the environment for change is different from the last attempt in the early 1990s at the beginning of the Clinton administration.

A bipartisan effort is underway to delineate the basic principles that would be necessary to undertake a large legislative reform. This effort will aim to control rising healthcare costs while expanding coverage for the 47 million uninsured in America, reforming the delivery system and figuring out how to pay for it.

Chronic diseases (such as COPD) consume large amounts of healthcare dollars and chronic disease advocates will need to ensure that the discussion includes quality and value goals such as prevention and evidence based measures.

We expect legislation that languished in the 110th Congress to be reintroduced in the next session. These bills include:

- Medigap parity for those on disability in all 50 states
- Elimination of the two-year waiting period for SSI disability
- Eliminating lifetime insurance caps
- Medicare Part B reimbursement for respiratory therapists

That being said, I would like to recap for you the successes we had in 2008 and the impact of these measures on the Alpha-1 Community:

Genetic Information Nondiscrimination Act

As you know, GINA passed and regulations to implement genetic nondiscrimination are being drafted. The passage of GINA prompted the Alpha-1 Foundation to pose the question of shifting from targeted detection to population screening. A workshop was held in the fall which will result in policy recommendations and pilot studies to explore the questions around newborn screening and aggressive family testing.

Pulmonary Rehabilitation

The Medicare bill that passed in the summer of 2008 includes the authority for the Centers for Medicare and Medicaid Services (CMS) to make pulmonary rehab a permanent Medicare benefit. The coding and billing of this service will help to define who is eligible for this benefit and which healthcare professionals will provide it. The Alpha-1 Foundation attended a meeting with CMS at the invitation of a coalition of healthcare provider groups to determine the outstanding issues for CMS to implement this benefit in January of 2010.

Air travel with supplemental oxygen

The Department of Transportation (DOT) published the amendment to the Air Carrier Access Act making it mandatory for all airlines doing business in the US to allow passengers to carry aboard their own FAA-approved portable oxygen concentrators by the spring of 2009. It is very important to note that the amendment gives individuals enforceability if they are discriminated against. The DOT did not address the issue of mandatory oxygen being provided for passengers who require it. The DOT is expected to issue another rule to address this question in early 2009, allowing time for public comment.

Home Oxygen Reimbursement for Medicare Beneficiaries

Congressional action last summer retained the 36-month payment cap for home oxygen, but changed the transfer of ownership from the beneficiary to the home care provider (durable medical equipment company). This means that after 36 months, individuals will no longer have a co-pay, but providers who have been receiving uncapped payments will now still have to continue to provide service. Individuals should make sure that their oxygen prescriptions are up to date and call 1-800-MEDICARE if you are concerned about changes in your home oxygen service.

We would love for each of you to become involved in our efforts and encourage you to follow these issues in the media. The Alpha-1 Foundation and Alpha-1 Association urge you to contact your senators and representatives and let them know that these issues are important to you and your loved ones. Visit the House of Representatives at www.house.gov to learn how to contact your representative, or visit www.senate.gov to learn how to contact your senator. You may also call the Capitol switchboard at 202-225-3121 to be directed to your legislator.

Ask the Alpha Doc

By Charlie Strange, MD
Professor of Pulmonary, Critical Care, Allergy and Sleep Medicine

Q. Is there a connection between Alpha-1 and fibromyalgia?

A. Fibromyalgia is a debilitating disease in which pain appears throughout the muscles of the body. Often there are trigger points (commonly along the neck, shoulders, and back) that reproduce the pain when stimulated. The cause of fibromyalgia is unknown, but it may be worsened by a variety of factors, including stress, sleep deprivation, and worsening of other diseases. For these reasons, fibromyalgia is seen accompanying many diseases. Pain control is not easy to obtain in this disease.

So, do those with Alpha-1 have an increased likelihood of suffering from fibromyalgia? The answer remains unknown.

Is there stress, sleep deprivation and severe lung and liver disease in Alpha-1? Absolutely!

Therefore, one of the answers to control of pain in Alpha-1 is to de-stress, get enough high quality sleep, and adequately treat the lung and liver disease when present.

Fibromyalgia experts are difficult to find but are available, usually in the Rheumatology community of physicians. Rheumatologists see patients with arthritis and are well versed in this difficult disease.

Expert panel suggests pilot studies on newborn screening for Alpha-1

By Dawn McGee, MS, CGC

EXPERTS IN ALPHA-1, newborn screening, and public policy met in Arlington, VA, in September to discuss whether newborn screening is appropriate for Alpha-1 Antitrypsin Deficiency.

The meeting was in response to the recent passage of the Genetic Information Nondiscrimination Act (GINA) into law in May, 2008. GINA provides protection from genetic discrimination in regard to health insurance and employment. The health insurance protections will become effective May 21, 2009 – 12 months after the signing of the bill. The employment protections will become effective 18 months after the bill's signing: Nov. 21, 2009.

In terms of health insurance protection, GINA prohibits using genetic information in determining eligibility or premiums by group and individual health insurers. The law also prohibits an insurer from requiring or requesting a genetic test.

GINA also prohibits an employer from using genetic information in making employment decisions – an employer or potential employer cannot use genetic information in the decision to hire, fire, or promote an employee. GINA also prohibits requiring, requesting, or purchasing genetic information about an individual or family member by an employer.

While GINA provides some much-needed protection from genetic discrimination, there are many limitations. GINA only provides protection for health insurance; it does not include life, disability, or long-term care insurance.

Also, while GINA forbids health insurers from discriminating based on genetic information alone, it does not prohibit insurers from underwriting based on current health conditions. For example, it would forbid a health insurer from refusing coverage due to someone's Alpha-1 genes, but the insurer could refuse coverage based on pre-existing COPD – even though Alpha-1 was the cause of the COPD.

GINA also does not require a health insurance company to cover any particular test or treatment, and does not prohibit the recommendation of genetic tests by health care professionals. In addition, GINA is not applicable to military personnel.

The potential benefits of newborn screening for Alpha-1 include both immediate medical intervention and the avoidance of risk factors to prevent future damage.

Newborn screening began over 40 years ago as a public health program regulated at the state level; so any newborn screening program and the conditions that are screened for are determined on a state-by-state basis.

Typical criteria for a condition to be appropriate for newborn screening:

There must be an available treatment; early initiation of treatment prior to onset of symptoms should significantly reduce, if not eliminate, the severity of the condition; the condition would not be detected by routine physical exam of a newborn; screening must be cost-effective; and screening should have essentially no false-negative and relatively few false-positive results. In addition, states need the infrastructure in place to screen every newborn, provide follow-up testing when indicated, and provide appropriate follow-up to physicians and families.

The work of Robert Guthrie, MD led to the creation of newborn screening. In the 1960s, Guthrie developed a way to screen for Phenylketonuria (PKU) by using blood drops collected on filter paper. This made cost-effective newborn screening possible. Babies diagnosed with PKU are given a specialized diet that prevents any of the clinical features of PKU from developing. Over time, states began to add to the list of health conditions for which all newborns are screened at birth. The procedure involves a heel stick and collecting the drops of blood on filter paper.

In 2006, the American College of Medical Genetics (ACMG) published recommendations for a core panel of conditions for which all states should screen. Specifically, the ACMG guidelines listed 29 core conditions and 25 secondary conditions as a uniform panel that should be required in all states. This helps to standardize state newborn screening programs; however, Alpha-1 was not listed as one of the core or secondary conditions.

The expert group that convened in Arlington in September was charged with determining, based on current standards, if Alpha-1 Antitrypsin Deficiency met the criteria to be added to the panel of conditions for which all newborns are screened.

The attendees reviewed the guidelines for newborn screening, the current screening methods for newborns, and current Alpha-1 testing methods. The benefits of early detection for Alpha-1 and the patient perspective were presented, and the panel discussed the two previous screening programs for Alpha-1, one in Oregon and one in Sweden. Finally, the experience of adding Cystic Fibrosis to the panel for newborn screening was presented. The attendees separated into smaller working groups to address the policy concerns, rationale, and feasibility of newborn screening for Alpha-1.

The consensus of the group was that multiple pilot studies need to be performed, due to the limited information available regarding the benefits of newborn screening for Alpha-1.

The potential benefits of newborn screening for Alpha-1 include both immediate medical intervention and the avoidance of risk factors, such as cigarette smoke and environmental exposures. This meeting illustrates that newborn screening is a new priority for the Alpha-1 community and more research and information on this topic is on the horizon.

Featured Clinical Resource Center

Loyola University

By Daniel Dilling, MD

LOYOLA UNIVERSITY HOSPITAL is delighted to have been asked to present our approach to Alpha-1 care in this newsletter. We offer a full range of treatment options, from watchful waiting and monitoring, to COPD care, to lung transplantation. We also offer treatment options for liver disease and panniculitis.

Daniel Dilling, MD, a pulmonologist and Alpha-1 specialist at Loyola, does not like to rush patients through appointments. "I believe strongly in listening to patients and giving them a lot of time, rather than just looking at test results and moving to the next patient," he said.

Dilling and other specialists at Loyola also offer long-term continuity of care. A patient may see the same doctor from the time of diagnosis through a progression of treatments, up to and including a lung transplant if necessary.

Loyola's lung transplant program is the largest and most successful in Illinois. Loyola has been doing lung transplants since 1988, and does more than 30 per year. The three-year survival rate is about 10 percent higher than the national average. Through our history, we have performed lung transplantation on a total of 38 patients with end-stage COPD from Alpha-1 Antitrypsin Deficiency – including eight in the past five years.

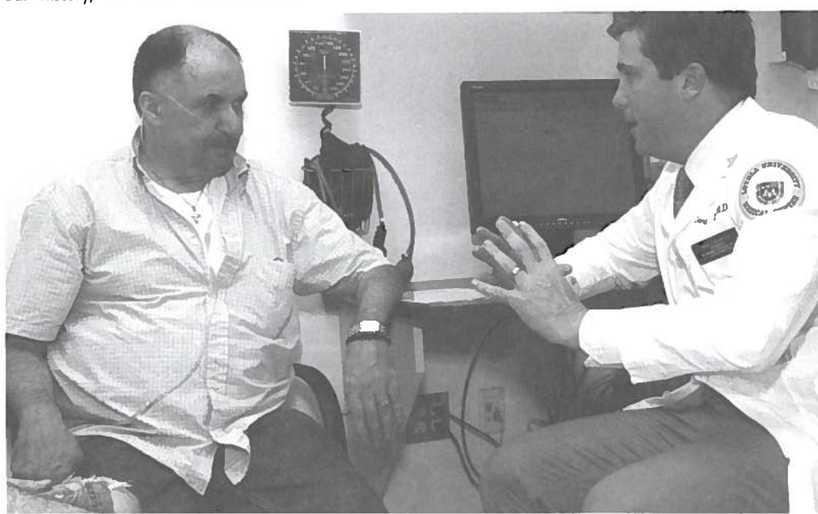
Loyola researchers are studying how to minimize the problem of chronic rejection in lung transplant patients. In one approach, they are investigating whether administering a certain structural protein could train the immune system to accept a transplanted lung, rather than attack it.

Loyola University Hospital is in west suburban Maywood, 13 miles west of the Chicago Loop and eight miles east of Oak Brook.

Loyola's treatment approach includes early consultation with a specialist and proper use of inhaler medication. Dilling, our Alpha-1 specialist, prescribes augmentation therapy only when appropriate. (He has no financial relationship with, and accepts no compensation from, any biopharmaceutical company). Dilling is board-certified in pulmonary disease, critical care, internal medicine and sleep medicine.

Other Loyola specialists who treat Alpha-1 patients include pulmonologist Patrick Fahey, MD, Medical Director, Respiratory Care; and liver specialist Claus Fimmel, MD, Medical Director, Hepatology. Fahey is board-certified in pulmonary disease and internal medicine; Fimmel is board-certified in transplant hepatology, internal medicine and gastroenterology.

Alpha-1 nurse coordinator Karen Pelletiere, RN, is available for information or new appointments by calling **708-327-5864** or faxing 708-327-2424.



CONSULTATION
Daniel Dilling, MD, talks with Alpha-1 patient Mike Kladis of Riverside, IL.

Featured Clinical Resource Center

Northwestern University COPD Program and Alpha-1 Clinical Resource Center

By Ravi Kalhan, MD, MS

THE NORTHWESTERN UNIVERSITY COPD PROGRAM and Alpha-1-Antitrypsin Deficiency Clinical Resource Center have the overall goals of providing outstanding clinical care, patient education and outreach, and state-of-the-art research into obstructive airway disease.

Our team consists of Ravi Kalhan, MD, director of the COPD Program at Northwestern, Linda Muszynski, RN, clinical pulmonary nurse, and Michelle Morley, clinical research coordinator for the COPD Program. In addition, physicians-in-training provide clinical care and conduct research under the auspices of our program.

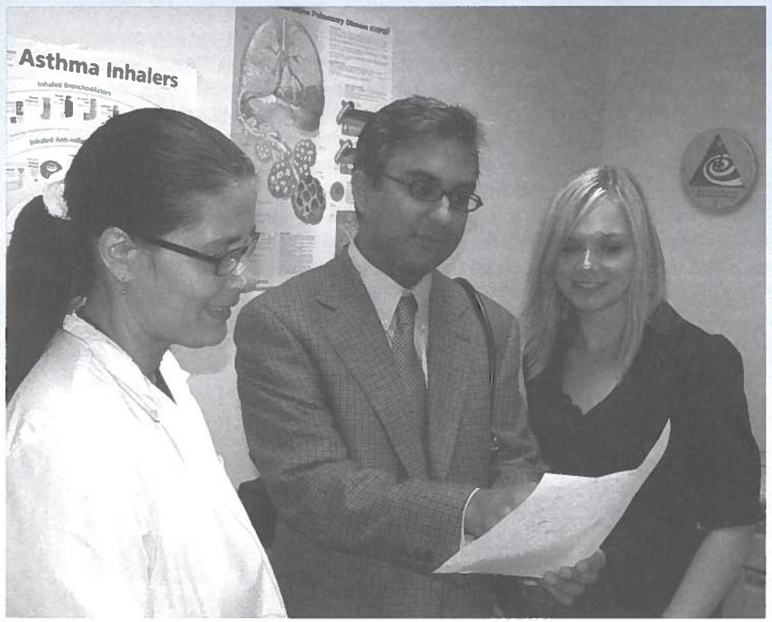
Working with the Alpha-1 Foundation builds on the mission of the Northwestern COPD Program: to work with our patients to further understanding of COPD and available therapies by providing education, coordinated medical care, and opportunities for research.

Our pulmonary rehabilitation program helps patients to better recognize and control breathing symptoms and improve breathing through education, supervised exercise, breathing technique training, nutritional counseling, and psychosocial support. Our pulmonary rehabilitation program provides services to 16 patients every two months.

We also work with the community on smoking cessation, COPD awareness, and resources for patients living with COPD. We conduct quarterly "spirometry days" in the metropolitan Chicago area to provide screening and enhance awareness regarding lung disease, and twice a year we conduct a large COPD patient education event on the Northwestern campus. For patients with liver disease, we work closely with faculty in Northwestern's Division of Hepatology and the Kovler Organ Transplantation Center at Northwestern Memorial Hospital.

Research is another key component of Northwestern's COPD Program, providing us with the opportunity to help not only our patients, but also the patients that we may never meet.

Our principal areas of interest include trials of novel therapies for COPD, including minimally-invasive approaches to lung volume reduction in emphysema, as well as other pharmacotherapies; investigations into the mechanisms of poor sleep quality in COPD; and expanding our knowledge regarding markers of early COPD and ways to interrupt the disease process early in its course.



COPD CONVERSATION Ravi Kalhan, MD, MS, director of the Northwestern University COPD program, with Linda Muszynski, RN, at left, pulmonary nurse coordinator, and Michelle Morley, clinical research coordinator.

To contact us for questions about the program or to request an appointment:
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Contact Person: Linda Muszynski, RN, Clinical Nurse
Email: **Linda.Muszynski@nmff.org**
Tel: 312-695-1670
Fax: 312-695-4741

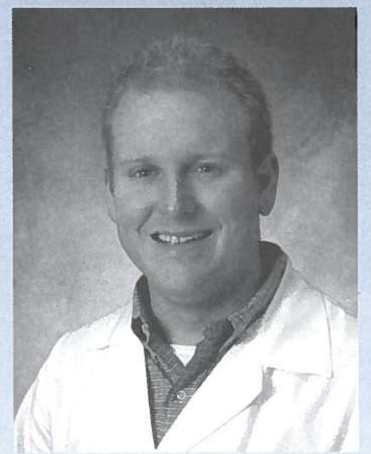
Featured Clinical Resource Center

Alpha-1 Program at University of Chicago Medical Center

By D. Kyle Hogarth, MD, FCCP
Assistant Professor of Medicine, University of Chicago Medical Center

In 2005, the Alpha-1 Antitrypsin Deficiency Clinical Resource Center (CRC) at the University of Chicago Medical Center became the first Alpha-1 Foundation approved CRC in Illinois. This distinction recognizes the University of Chicago's comprehensive approach and focused expertise in addressing the many dimensions of this condition, including the Medical Center's ability to treat advanced liver and lung damage and its experience in liver and lung transplantation. I am the medical director of the program, and I am actively involved in Alpha-1 research and advocacy.

THE UNIVERSITY OF CHICAGO program attracts patients from throughout the Midwest and as far away as Alaska. Many patients come here for initial treatment planning, stabilization of their secondary complications such as COPD, patient education, and for treatment of advanced complications. To minimize long-term disruption to one's life, our Alpha-1 team will coordinate with each patient's local physician for ongoing, day-to-day medical management. This arrangement enables patients to balance the multidisciplinary expertise of the academic medical center with the convenience of receiving routine medical care close to home.



Treatment at the University of Chicago Medical Center begins with an extended consultation with me (I am board certified in pulmonary and critical care medicine as well as internal medicine). Each patient (and family, if desired) can expect to spend at least two hours of dedicated time meeting and talking with me at their first visit. This is a very helpful opportunity for the patient and family to understand the diagnosis, ask many questions and address concerns. Subsequent consultations are provided on an ongoing basis as medical needs change or questions arise.

Some individuals with advanced disease eventually require a lung or liver transplant to replace the severely damaged organ. The University of Chicago Medical Center is one of the leading transplant centers in the US.

Patients needing transplant benefit from our depth of experience, innovation and expertise. The University of Chicago Medical Center has the fourth oldest liver transplant program in the US and a lung transplant program directed by one of the nation's earliest pioneers in lung transplantation. Both areas have more than 20 years of experience.

In addition to performing single-organ cadaveric transplants, the University of Chicago team has been successful with complex, multiple-organ transplants. This medical center also has been an innovator in organ transplantation, including the first living-donor liver transplant in the US and the first successful split-liver cadaver transplant in the US.

Patients requiring liver or lung transplantation receive comprehensive medical, educational and emotional support before and after surgery, as well as lifelong monitoring to regulate immunosuppression and address issues of organ rejection or other potential complications.

In addition to medical care directed by physicians, patients may benefit from additional services provided here, including:

- Smoking cessation classes and counseling
- Genetic counseling to address the inherited aspect of this disorder
- Comprehensive pulmonary rehabilitation to increase lung function and overall energy level
- Nutrition services provided by registered dietitians, to address nutrient deficiencies and develop guidelines for healthy eating
- Patient education, an important component for lifelong management of Alpha-1

The Alpha-1 clinic at the University of Chicago can be reached by telephone at 773-702-9660 and fax at 773-834-7068. My email address is **dhogarth@uchicago.edu**.

Take the Celtic Challenge – or pick your own challenge!

By Angela McBride
Alpha-1 Foundation, Development Director

THE ALPHA-1 FOUNDATION was founded on the idea that individuals can make a difference. Underlying the achievements of the Alpha-1 community is the conviction that change can come if everyone does his or her part to make it happen.

And that's the point of the Alpha-1 Foundation's **Building Friends for a Cure** (BFC) program. Since a central part of the Foundation's mission is funding research to improve health and ultimately find a cure for Alpha-1, we want to keep research going strong.

We had a great year in 2008. To make 2009 even more successful, we need YOUR help! And we're convinced that everyone can help in their own way.

Would you like an example?

Well, why not accept the Celtic Challenge?

What is the Celtic Challenge?

It's celebrating your Irish heritage and raising funds for a cure for Alpha-1.

How do you do that? Easy… you party! Organize a St. Patrick's Day party. Invite your family and friends to be a part of the cure all while having a good time.

If you are interested in having a good time for a good cause, let us know. The Alpha-1 Foundation staff is here to help you get started. We can help you brainstorm, provide suggestions and put you in touch with volunteers who have organized a similar event.

What, you're not Irish? Come on. We think everyone is Irish on St. Patrick's Day!

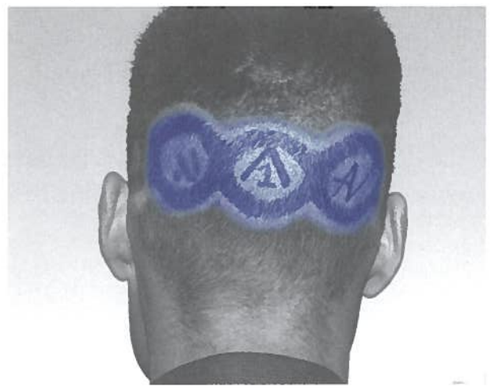
But just suppose St. Patrick is not your kind of guy – or organizing a party isn't your thing. Do you have another idea? It doesn't have to be anything big. Make it something that interests you – that you'd enjoy; and remember to have fun!

Need some help? Just ask. To get you started, we'll provide you with our Building Friends for a Cure Manual. It includes fundraising guidelines and a list of fundraising ideas that have been proven successful.

You can also check the calendar at the end of this article, for events already scheduled. We think you'll find them interesting and maybe even inspiring. Besides the Celtic Challenge, there are two "**Jeans for Alpha-1 Genes**" programs (an event in schools, where teachers, and sometimes students, are allowed to pay to wear jeans to school. The money raised will go towards Alpha-1 research.) There's also the second annual **George Washington Bridge Walk** in New York and New Jersey, May 9; and the **Get the Scoop on Alpha-1 Ice Cream Event** in Denver, CO, May 15.

Here's another possibility. Join our **Firstgiving online fundraising campaign** at http://www.firstgiving.com/alpha-1foundation. The website makes it easy to create your own personal web page. Perhaps the easiest way to begin is to join our **letter-writing campaign** – you don't have to organize an event from beginning to end. You can join many other Alphas who send letters to raise money and awareness for Alpha-1. You direct friends to your personal web page, where they can make a donation right online if they wish. Letter-writing campaigns are successful because friends are asking friends to support them in their fundraising efforts.

HAP AND DIANE EATON took a break in Miami during their incredible 10,000-mile trek, pedaling a tandem bicycle across the country to raise awareness and funds for Alpha-1.



RICHARD O'BRIEN — Team Alpha-1's "sweeper" at the annual ALA of Massachusetts Escape to the Cape bike trek — always gets this "hair tattoo" just before the big day. Below, Team Alpha-1 is presented the Silver Spoke Award for having the most riders in the event.

If you'd like more inspiration, here are some of our successes in 2008:

- The "Half Shamrock Marathon 8K Race" in Virginia Beach, VA, in March, spearheaded by Jennifer Clark.
- In May, a flood of events, including the First NY/NJ George Washington Bridge Walk; the Foundation's Internet Mother's Day Scarf and Card Campaign; and the Alpha Okies Silver Horn Golf Tournament, Dennis Pollock, his support group, family and friends.
- In June, Frank Deford was master of ceremonies at the Breath of Life Cocktail Reception in Greenwich, CT, organized by Ken and Bettina Irvine. Our Internet Father's Day Tie and Card Campaign was June 15, and Karen Erickson led a group of friends in the Breathe Easy Bike Ride in Santa Inez, CA.
- Sheila Favazza, Susan Binnall, and the Massachusetts support group arranged the Plymouth Harbor Cruise July 26.
- In August, Ed Mikell, who has two grandsons with Alpha-1, worked to publicize the English Channel swim by five of his friends on the New York City Alpha-1 Swim Team. They drew media coverage on both sides of the Atlantic and raised more than $11,000 for Alpha-1 research.
- Lou Glenn and Jennifer Jacks led the Lone Star Alphas Shoot for a Cure golf tournament in Flower Mound, TX, Oct. 6.
- Team Alpha-1 had its biggest year ever at the ALA of Massachusetts Autumn Bike Trek – the annual "Escape to the Cape." Team Alpha-1 far surpassed its fundraising goal, and was awarded the first "Silver Spoke Award" for having the most riders involved. Team Alpha-1's participation in the event was organized by the large group known as the East Coast Alpha Friends & Family.

Below is the calendar of events scheduled in the first half of 2009. You can be sure we'll have lots more events as the year goes on.

Building Friends for a Cure
2009 Calendar of Upcoming Events

When?	What?	Where?	Who?
March 13	Celtic Connection Dinner & entertainment	Braintree, MA	Sue Binnall **sbinnall@comcast.net**
March 17	Celtic Challenge (Please contact the Alpha-1 Foundation for more information)	USA	Yiomara Perry **yperry@alphaone.org**
March 30	Jeans for Alpha-1 Genes	Mansfield, MA	Donna Tucker **hdtuck4@yahoo.com**
May 1	Jeans for Alpha-1 Genes	Montclair, NJ	Nancy Smith **njsmith1029@aol.com**
May 9	New York & New Jersey George Washington Bridge Walk	New York, NY	Joe Reidy **joereidy@verizon.net**
May 15	Get the Scoop on Alpha-1 Ice-Cream Event	Denver, CO	Judy Simon **saidsimon@comcast.net**
June 8	NYC Liver Walk at Battery Park	New York City	Rose McClellan **McClellanR@sullcrom.com**
June 27	Lung Association of Southern California Breathe Easy Ride	Santa Ines, CA	Karen Erickson **kerick17@hotmail.com**

BETTINA IRVINE presents the "Alpha-1 Viking Explorers Award" to New York City Fire Department Chief Medical Officer David Prezant, MD, at the Breath of Life cocktail reception organized by Bettina and her husband Ken. Frank Deford hosted the reception in Greenwich, CT.

The goal of the Alpha-1 Foundation's Building Friends for a Cure Program is to build stronger links between the organization and the Alpha-1 community using social events to increase awareness and raise funds for Alpha-1 research and programs. For information, contact Yiomara Perry at 888-825-7421, Ext. 248 or yperry@alphaone.org

Team Alpha-1 was created to encourage participation of Alphas in athletic events to promote awareness and raise funds in support of research and programs. The sponsors are Baxter Healthcare, CSL Behring and Talecris Biotherapeutics.

2009 National Education Programs

Education Days, Events and Meetings

The following calendar features a partial list of events. For more current listings, check the website at **www.alphaone.org.**

DATE	EVENT	LOCATION
March 21, 2009	**Alpha-1 Education Day & Advocacy Training**	Los Angeles, CA
April 18, 2009	**Alpha-1 Education Day**	Jacksonville, FL
August 8, 2009	**Alpha-1 Education Day**	Denver, CO
August 29, 2009	**Alpha-1 Education Day**	Hershey, PA
November 14, 2009	**Alpha-1 Education Day**	Ann Arbor, MI
TBA	**Alpha-1 Education Day**	Boston, MA
June 5-7, 2009	**Alpha-1 Association National Conference**	San Francisco, CA

The Alpha-1 National Education Series is co-sponsored with the Alpha-1 Foundation and is made possible by unrestricted educational grants from **AlphaNet, Centric Health Resources, CSL Behring, Baxter** and **Talecris Biotherapeutics**. For information on attending or exhibiting at an education program, contact Marlene Erven at 1-800-521-3025 or email mserven@alpha1.org.

*Commitments and dates are subject to change.

Alpha-1 Foundation

The Alpha-1 Foundation is a not-for-profit organization dedicated to providing the leadership and resources that will result in increased research, improved health, worldwide detection, and a cure for Alpha-1 Antitrypsin Deficiency (Alpha-1). The Foundation has invested $35 million to support Alpha-1 Antitrypsin (AAT) research and programs in nearly 70 institutions in North America and Europe.

Alpha-1 Association

The Alpha-1 Association is a member-based not-for-profit organization founded in 1991 to identify those affected by Alpha-1 Antitrypsin Deficiency and to improve the quality of their lives through support, education and advocacy. The Association has a network of 75 volunteer-led support groups around the U.S.

AlphaNet

AlphaNet, Inc. is a unique disease management organization. Through its medical and operations staff, AlphaNet provides a wide range of integrated support services to individuals with Alpha-1 Antitrypsin Deficiency who require augmentation therapy, oversees and sponsors clinical trials involving Alpha-1 therapies, and makes available a comprehensive disease management and prevention program to improve the quality of life of those affected by Alpha-1.

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