Identifying Alpha 1-Antitrypsin Deficiency: an Under-Recognized Cause of Pulmonary Disease

James K. Stoller, M.D., M.S.

Alpha 1-antitrypsin deficiency is an under-recognized cause of chronic obstructive pulmonary disease. Of the 100,000 Americans affected by this deficiency, fewer than 10 percent are clinically recognized.

Data from a 1994 study of readers of a national newsletter by Stoller et al, indicate a mean interval delay of 7.2 years between initial symptoms of alpha 1-antitrypsin (AAT) deficiency and first diagnosis. In the study, 43 percent of respondents with severe AAT deficiency reported seeing at least three physicians before the initial diagnosis was made, and the delay in diagnosis was associated with adverse psychosocial effects.

Alpha 1-antitrypsin deficiency has been a special interest of Cleveland Clinic pulmonary clinicians since the mid-80s. During that time, The Cleveland Clinic served as the Data Coordinating Center for the National Heart, Blood and Lung Institute Registry for Individuals with Severe Deficiency of Alpha 1-Antitrypsin. The Registry was a multi-center study (37 centers), which assessed the natural history of AAT deficiency based on observations of 1,129 individuals followed for up to seven years.
ENHANCING SUSPICION

In view of the clinical under-recognition of AAT deficiency, there is a clear need to enhance suspicion to prompt effective treatment. For example, in individuals with moderately severe obstructive lung disease, observational data from European studies and from the NHLBI Registry indicate that recipients of intravenous augmentation therapy experienced a slower rate of decline in lung function than did non-recipients.

Additionally, because AAT deficiency is a genetic disease, recognition of a severely deficient individual might prompt genetic counseling and testing of family members.

Because of the Clinic’s long-standing interest and expertise in AAT deficiency, an Alpha 1-antitrypsin Deficiency Center of Excellence has been established. Supported by a grant from Joseph Bennett and Family and the Alpha-1 Foundation, the center offers specialty care to patients, an ongoing patient registry, patient and family education (including the annual Jean Wall Bennett Conference on Alpha 1-antitrypsin Deficiency), and continued research into treatment.

INVESTIGATING GENETIC MODIFIERS

Recent research activities include the investigation of a new pooled human plasma of alpha 1-antitrypsin protein for augmentation therapy and participation in studies evaluating inhaled transgenic alpha 1-antitrypsin. In collaboration with Harvard University investigators, Clinic researchers currently are studying genetic modifiers of lung disease in individuals with severe alpha 1-antitrypsin deficiency, type PI*ZZ.

For the practicing pulmonologist confronted with a patient with AAT deficiency, the Cleveland Clinic serves as a national referral center for clinical management, from conventional COPD management and augmentation therapy to participation in novel therapeutic approaches. Patients can participate in the Cleveland Clinic’s registry and gain access to resources through the Alpha-1 Foundation and the Alpha-1 Association.

For more information, or to refer a patient, contact Dr. Stoller at 216/444-1960 or 800/553-5056, ext. 41960.
**Clinical Features Suggestive of Alpha 1-Antitrypsin Deficiency**

- Early-onset emphysema (i.e., younger than age 50)
- Emphysema and liver disease
- Emphysema with a basilar distribution of hyperlucency on chest X-ray or CT scan
- Emphysema in a minimal smoker or person who has never smoked
- Family history of emphysema
- A young person with airflow obstruction that fails to normalize with maximal bronchodilator therapy
- Panniculitis
- Bronchiectasis without evident etiology
- Cirrhosis without evident etiology

Cuts from a chest CT scan of a patient with PI*ZZ alpha 1-antitrypsin deficiency show the characteristic basilar hyperlucency. Emphysematous change is illustrated in the apical cut (above left), but is more pronounced in the basal cut (above).

**Recent Publications**

Investigators from the Cleveland Clinic’s departments of Pulmonary, Allergy and Critical Care Medicine, and Hematology and Oncology will be enrolling patients with systemic mastocytosis in a pilot trial to study the safety and efficacy of imatinib mesylate (STI-571 or Gleevec).

The ultimate purpose of the trial, which is being led by Fred H. Hsieh, M.D., and Clinic oncologist Alan Lichtin, M.D., is to offer a therapeutic option to patients with high-grade systemic mastocytosis, where the potential benefits of reducing the body’s mast cell burden outweigh the risks of Gleevec therapy.

The mast cell is a vital effector cell of the immune system and plays a key role in the pathogenesis of asthma and allergic inflammation. It also has the primary role in the pathogenesis of systemic mastocytosis.

Patients with systemic mastocytosis have abnormal mast cells in the bone marrow as well as other organs, including the skin, lymphatic system, liver, spleen, stomach and intestine, bone, CNS and vascular system. (Patients with cutaneous mastocytosis have characteristic skin lesions without evidence of systemic involvement.) The disease may be low-grade and indolent or may be high-grade and associated with malignancies or other hematologic disorders.

**IMPROVING THE STANDARD OF CARE**

The current standard of care involves medical management to provide symptom relief, which is modestly effective at best. There are no FDA-approved therapies available to target the survival and proliferation of the mast cell. Only an identified malignancy is treatable with standard chemotherapy.

The Gleevec trial will be limited to adult patients with high-grade disease, confirmed by bone-marrow biopsy, and identified mutations in their c-kit gene.

Gleevec is one of the first therapeutics rationally designed to inhibit receptor tyrosine kinase activity. The drug is FDA-approved to treat patients with chronic myeloid leukemia by targeting the BCR-ABL oncogene and to treat gastrointestinal stromal tumors by targeting the mutant c-kit oncogene. The c-kit mutations are linked to the pathogenesis of systemic mastocytosis, because c-kit provides essential intracellular survival signals for the mast cell.

The Gleevec trial is an open-label, unblinded, single-arm study of patients with bone marrow biopsy-proven systemic mastocytosis syndromes. Individuals with smoldering systemic mastocytosis, systemic mastocytosis with an associated hematologic non-mast cell disease, aggressive systemic mastocytosis, mast cell leukemia and mast cell sarcoma will be considered. All patients will undergo genetic analysis of their c-kit gene prior to enrollment; only patients with identified c-kit mutations will be enrolled.

**TRIAL ENDPOINTS**

Clinical trial participants will receive six months of active drug therapy. The primary endpoint is reduction in tissue mast cell load. Secondary endpoints are reduction in chemical markers of mast cell load and symptom scores.

For more information, or to refer patients for the trial, contact Dr. Hsieh at 216/444-3504 or 800/553-5056, ext. 43504.

Note: This research is supported by Novartis Pharmaceuticals.
Cleveland Clinic pulmonologist Raed A. Dweik, M.D., was granted a $633,000 K23 award from the NIH, which will allow him to study nitric oxide and carbon monoxide in patients with pulmonary disease.

NIH “K” Awards Help Launch Physician-Investigators

Physicians in the Department of Pulmonary, Allergy and Critical Care Medicine are heavily involved in basic and clinical research, the vast majority of which is aimed at finding better or viable treatments for lung diseases. Recent K awards from the National Institutes of Health/National Heart, Lung, and Blood Institute include:

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<tr>
<th>GRANT</th>
<th>PRINCIPAL INVESTIGATOR</th>
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<tr>
<td>Mid-career award in patient-oriented research (K24) to provide resources for an established investigator to mentor young trainees</td>
<td>Serpil C. Erzurum, M.D.</td>
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<tr>
<td>Mentored clinical scientist development award (K08) to study signaling and allergic lung disease: NF-kB&amp;T helper subsets</td>
<td>Mark A. Aronica, M.D.</td>
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<tr>
<td>Mentored patient-oriented research career development award (K23) to study nitric oxide and carbon monoxide in pulmonary hypertension</td>
<td>Raed A. Dweik, M.D.</td>
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<tr>
<td>Mentored clinical scientist development award (K08) to study effector mechanisms in allergic inflammation</td>
<td>Fred H. Hsieh, M.D.</td>
</tr>
<tr>
<td>Patient-oriented research career development award (K23) to study GM-CSF’s role in macrophage surfactant catabolism</td>
<td>Tracey L. Bonfield, Ph.D.</td>
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The project’s content is offered free as a service of The Cleveland Clinic Center for Continuing Education.

Clinic Launches Online Medical Reference Guide

Access up-to-date disease information and practical treatment strategies — at no charge — through The Cleveland Clinic Disease Management Project at www.clevelandclinicmeded.com/diseasemanagement. Each chapter is designed to incorporate national clinical practice guidelines in a simple, straightforward fashion. If an author disagrees with the guidelines, they indicate as such and include their reasons.

The section on Pulmonary Disease contains the following chapters:

- Asthma
- Chronic Obstructive Pulmonary Disease
- Community-acquired Pneumonia
- Cough
- Hospital-acquired Pneumonia
- Idiopathic Interstitial Lung Disease
- Lung Cancer
- Pleural Disease
- Upper Respiratory Tract Infections

In addition to pulmonary disease, the site features sections on cardiology, dermatology, endocrinology, gastroenterology & hepatology, hematology & oncology, infectious disease, nephrology, neurology, psychiatry & psychology, and women’s health.

The project’s content is offered free as a service of The Cleveland Clinic Center for Continuing Education.
Chronic beryllium disease (CBD or berylliosis) is an occupationally acquired granulomatous lung disease similar to sarcoidosis and caused by exposure to beryllium. About 2 percent to 10 percent of exposed individuals will develop beryllium hypersensitivity, in which there is proliferation of beryllium-specific T cells. One percent to 5 percent of exposed individuals will develop CBD.

**OCCUPATIONAL EXPOSURE**

The most significant exposure is in the occupational setting, particularly industries related to primary production, metal machining and reclaiming scrap alloys. However, as the use and application of beryllium and its alloys expand, so does the risk of exposure.

Despite the advent of industrial exposure control measures to minimize air levels, CBD continues to occur. The Cleveland Clinic remains on the forefront in the screening of individuals exposed to beryllium and the evaluation and management of individuals who become sensitized to beryllium or develop CBD.

**A GENETIC LINK**

In addition to environmental exposure, genetic predisposition seems to have a major role in the development of CBD. A variant of the human leukocyte antigen [HLA-DPb1(Glu69)] is found in 80 percent to 97 percent of patients with CBD and only in 30 percent of controls. An ongoing collaboration between Cleveland Clinic pulmonologists — including Herbert Wiedemann, M.D., and Dr. Dweik — and Italy's Cesare Saltini, M.D., who was the first to describe this genetic association, has resulted in a better understanding of the role of genetics in CBD. A new collaboration with researchers at the National Institute of Occupational Safety and Health is focused on genetics and CBD.

**DIAGNOSING CBD**

The diagnosis of CBD is based on: (1) a history of beryllium exposure; (2) evidence of sensitization to beryllium by a positive blood or bronchoalveolar lavage beryllium-specific lymphocyte proliferation test (BeLPT); and (3) the presence of non-necrotizing granuloma on lung biopsy. If the history of exposure is subtle or unknown, sensitization to beryllium can be considered as evidence of exposure. Flexible fiberoptic bronchoscopy with bronchoalveolar lavage and transbronchial biopsy is usually necessary to confirm a suspected diagnosis of CBD.

The BeLPT is currently the screening test of choice to identify beryllium workers who develop beryllium sensitization or CBD. The Cleveland Clinic operates one of the five specialized laboratories in the country where BeLPT is performed.

Many similarities exist between CBD and sarcoidosis. All patients with sarcoidosis should undergo a detailed occupational history to rule out potential exposure to beryllium and the possibility of CBD.

**RESEARCH STUDY FINDINGS**

A Cleveland Clinic research team, led by Dr. Dweik, recently evaluated the relationship between smoking and CBD. Based on a study of more than 200 beryllium-sensitized workers, the preliminary findings revealed that smokers were at lower risk for developing CBD—even after controlling for the effects of age, sex and race. (In this regard, the “protective” effect of smoking on the development of CBD is similar to that which has been found for other granulomatous diseases, such as sarcoidosis and hypersensitivity pneumonitis, as reported by other investigators.) African-Americans, on the other hand, seem to be at increased risk for developing CBD.

In another study led by Dr. Dweik, individuals with CBD were found to have high levels of the inflammatory marker nitric oxide in their exhaled breath. These findings provide support for an association between active lung inflammation and CBD and provide a potential method for monitoring these patients. Dr. Dweik will continue to study nitric oxide and carbon monoxide in patients with pulmonary disease thanks to a $633,000
grant from the National Institutes of Health/National Heart, Lung and Blood Institute.

Currently there is no cure for CBD, and no controlled studies for CBD are available. However, based on anecdotal reports and the pathogenesis of the disease (immune mediated), and because of the similarities to sarcoidosis, CBD can be treated with corticosteroids. Since therapy is not curative and has significant side effects, it is only recommended for patients who are symptomatic or demonstrate decline in their pulmonary function. In patients who fail corticosteroids or develop significant side effects, methotrexate may be an option. Lung transplant may be considered in end-stage cases.

For clinical evaluation of suspected CBD or information about beryllium-induced lung disease, call 216/445-5763 or 800/553-5056, ext. 55763. To arrange for BeLPT testing, call 216/444-8844 or 800/553-5056, ext. 48844.

HISTORICAL SPOTLIGHT: HOWARD S. VAN ORDSTRAND, M.D.

In the early 1940s, Cleveland Clinic physician Howard S. Van Ordstrand, M.D., identified the first occupational beryllium disease case in acute form. He was known for his original description of acute berylliosis, detailing the association between beryllium exposure and lung toxicity in the first published U.S. cases of pneumonitis.

Dr. Van Ordstrand studied the disease with colleagues Sharad D. Deodhar, M.D., Ph.D., and Barbara P. Barna, Ph.D., Cleveland Clinic immunopathologists who demonstrated the immunological nature of berylliosis.

Dr. Van Ordstrand served as chairman of the Cleveland Clinic’s Division of Medicine, was the first head of the Cleveland Clinic’s Department of Pulmonary Disease and served a one-year term as president of the American College of Chest Physicians.

REFERENCE:

COMMON USES OF BERYLLIUM

Beryllium is a strong (stiffer than steel), light (lighter than aluminum) metal, with a high melting point and excellent thermal and electrical conductivity. Initially used in the nuclear weapons industry, its unique properties led to its use in many others, including: inertial guidance systems • turbine rotor blades • laser tubes • rocket engine liners • springs and gears • aircraft brakes and landing gear • ball bearings • injection and blow mold tooling • electrical contacts • automotive electronics • X-ray tube windows • spark plugs • electrical components • ceramic applications • aircraft engines • welding electrodes • computer electronics • golf clubs

RECENT PUBLICATIONS
Culver DA and Dweik RA.

Dweik RA.

Barna BP, Dweik RA, Farver CF, Culver D, Yen-Lieberman B, Thomassen MJ.


Samuel G, Wiedemann HP, Dweik RA.

Saber W and Dweik RA.

Dweik RA, Laskowski D, Erzurum SC.
Despite a Patient’s History of Allergy, Penicillin may be an Option

Mercedes Arroliga, M.D., and Alejandro Arroliga, M.D.

A history of allergy to penicillin does not necessarily rule out using penicillin again. With skin testing and, in some cases, desensitization, most patients with a history of penicillin allergy can safely receive the drug. In addition, by using fewer antibiotics in favor of penicillin and related antibiotics, it may be possible to reduce the number of multidrug-resistant bacteria.

**ALLERGIC REACTIONS TO PENICILLIN**

Because of its high efficacy and low toxicity, the penicillins are one of the most useful antimicrobial drugs. They are also frequently responsible for causing allergic reactions: Up to 25 percent of patients who are admitted to the hospital report allergies to antimicrobial agents, and penicillin is cited specifically most often.

Allergic reactions — mainly skin rashes, such as maculopapular or urticarial — are estimated to occur in approximately 2 percent of patients treated with penicillin. However, the most serious allergic reaction, penicillin-induced anaphylaxis, is life-threatening. According to some estimates, penicillin-induced anaphylaxis is responsible for 75 percent of anaphylactic deaths in the United States.

**PENICILLIN SKIN TESTING**

At The Cleveland Clinic, we have demonstrated that a penicillin skin test (PST) is a valuable tool for modifying antibiotic use in hospitalized patients. In a pilot study of 24 penicillin-allergic patients who were admitted to our medical ICU, 95 percent had negative reactions to a PST. In half of the patients (48 percent), a penicillin antimicrobial agent was started as a result of the negative skin test with no adverse effect.

The major breakdown products of penicillin are the penicilloyl group (about 95 percent), also known as the major determinant. Penicillin G, benzyl penilloate, benzyl penilloate, and benzyl propylamine are the so-called minor determinants (less than 5 percent). A PST can detect the presence of immunoglobulin E (Ig E) antibodies by using benzyl polylys in to test for the major determinants and penicillin G to test for minor determinants. The test can safely and reliably identify 97 percent of patients who are at risk for developing a severe immediate reaction to penicillin. (A mixture for minor determinants is not commercially available.)

**PREDICTING REACTIONS**

A history of penicillin allergy alone is not reliable in predicting a future immediate allergic reaction. It has been suggested that the presence of Ig E antibody to penicillin decreases with time, and up to 80 percent of the patients with a history of penicillin allergy will have an absence of Ig E antibody by 10 years. Therefore, the possibility of a new allergic reaction decreases with time. Our experience mirrors this thinking: In 92 percent of our patients, previous penicillin reactions occurred more than 10 years before they were given the PST.

**HALTING THE EMERGENCE OF MULTIDRUG-RESISTANT BACTERIA**

Typically, patients with a history of penicillin allergy are treated with alternative, non-beta-lactam antibiotics, such as vancomycin and quinolones. Their use has been associated with an increased rate of infections from multidrug-resistant bacteria, which contributes to higher morbidity, mortality and costs of care.

Programs designed to manage antimicrobial resistance perhaps should include modifications of antibiotic use, such as a reduction in the use of vancomycin, quinolones and third-generation cephalosporins, and an increased use of penicillins, with or without the use of aminoglycosides.

We recently reported a study of consecutive patients admitted to all of our ICUs with penicillin allergies noted in their charts. Only one of the 96 patients experienced a positive PST, from which no significant adverse event was reported. In those patients who were receiving antibiotics for therapeutic reasons, up to 82 percent were treated with a beta-lactam antibiotic.

Our experience suggests that a beta-lactam antibiotic can be used in most patients admitted to an ICU with a history of penicillin allergy. The PST can be used in hospital wards and outpatient areas. We believe that the PST is an underused, safe procedure that has the potential of modifying the pattern of antibiotic use in patients who report penicillin allergy. By reducing the use of antibiotics like vancomycin and the quinolones, we may be able to modify and decrease the emergence of multidrug-resistant bacteria.

For more information, or to refer patients, contact Dr. Mercedes Arroliga at 216/444-6933 or 800/553-5056, ext. 46933.

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**RECENT PUBLICATIONS**

Arroliga ME, Radojcic C, Gordon SM, Popovich MJ, Bashour CA, Melton AL, Arroliga AC.

Arroliga ME, Pien L.

For the 9th year in a row, U.S. News & World Report ranked The Cleveland Clinic as one of America's top hospitals. For the first time ever, The Cleveland Clinic is recognized for excellence in all medical specialties ranked by the magazine.

In addition, The Cleveland Clinic was ranked one of the top 10 respiratory centers in the country in the April 2003 issue of RT, the Journal for Respiratory Care Practitioners. The article acknowledged the Clinic’s more than 100 full-time respiratory care practitioners and the development of “therapist-driven protocols that use branched chain logic diagrams in creating respiratory plans.” The protocols have been used by hospitals throughout the country. James Stoller, M.D., medical director of Respiratory Therapy at The Cleveland Clinic, was quoted in the article. “Our protocol concept has been an outgrowth of our desire to come up with a better paradigm for developing the practice of respiratory care,” he said.

As head of the Pulmonary Vascular Disease Program of the Cleveland Clinic’s Department of Pulmonary, Allergy and Critical Care Medicine, Constance A. Jennings, M.D., is putting her energy into the research and treatment of pulmonary hypertension.

“Unlike many other pulmonary vascular disease programs, ours is managed by the Pulmonary Department. A multidisciplinary approach involves specialists from Cardiology in each patient’s evaluation, and consultations with physicians from Hepatology and Rheumatology are available as needed. Lung transplantation can be considered for disease that advances despite treatment. The Clinic is also one of a few centers nationwide to offer surgical treatment for patients with chronic thromboembolic pulmonary hypertension,” says Dr. Jennings.

Dr. Jennings earned her medical degree from Mayo Medical School in Rochester, and she completed her advanced training at Georgetown University Hospital in Washington, D.C. Following her fellowship, she spent several years in research and patient care, with a focus on interstitial lung disease at the NIH and at the National Jewish Center in Denver, Colo. Prior to joining The Cleveland Clinic, Dr. Jennings was a staff pulmonologist at Mayo Clinic.

Other members of the Clinic’s Pulmonary Vascular Disease Program include Alejandro Arroliga, M.D.; Jeffrey Chapman, M.D.; Atul Mehta, M.D.; and Omar Minai, M.D.

To refer patients to Dr. Jennings, call 216/445-4184 or 800/553-5056, ext. 54184.
Staying at the Forefront of Advanced Bronchoscopy

Updated Suite is Dedicated in Honor of Special Donors

A newly renovated bronchoscopy suite was dedicated in honor of Thomas P. and Patricia L. Brundige in July 2003. As part of the dedication event, members of the Pulmonary, Allergy and Critical Care Medicine Department gave updates on lung transplantation and current bronchoscopy-related research.

“With a state-of-the-art bronchoscopy facility, our physicians will be better able to manage our lung transplant patients,” says Atul C. Mehta, M.D., head of the section of Bronchology and Medical Director of Lung Transplantation. The advanced technology in the new suite will aid in the diagnosis of lung cancer, as well as the palliation of large airway obstruction.

The facility also will be great training ground for the Clinic’s pulmonary fellows, adds Dr. Mehta.

For further information, or to refer patients, call Dr. Mehta at 216/444-2911 or 800/553-5056, ext. 42911.

Cleveland Clinic Nurses Achieve Magnet Status

The Cleveland Clinic, including The Children’s Hospital, is one of only 72 hospitals throughout the entire United States to achieve Magnet status for excellence in nursing services by the American Nurse Credentialing Center. To earn Magnet status, a hospital must meet strict criteria based on various quality indicators and standards. Hospitals that achieve Magnet status are recognized for providing the highest standard of nursing care and excellence.

Areas of Expertise

Acute respiratory distress syndrome (ARDS)
Allergic rhinitis
Allergies (drug and food; latex)
Asthma
Beryllium-induced lung disease
Chronic obstructive pulmonary disease (COPD), including alpha1-antitrypsin deficiency
Interstitial lung disease
Interventional bronchology
Lung cancer
Lymphangioleiomyomatosis (LAM)
Pulmonary alveolar proteinosis (PAP)
Pulmonary vascular disease
Sarcoidosis
Sepsis
Sleep-disordered breathing
Urticaria
Weaning from mechanical ventilation

In collaboration with thoracic surgery colleagues, we evaluate patients for:

- Lung transplantation
- Lung-volume reduction surgery (LVRS) for emphysema
- Pulmonary thromboendarterectomy (for chronic pulmonary hypertension secondary to thromboemboli)

Referrals/Information

For more information, or to refer patients to the Cleveland Clinic’s Department of Pulmonary, Allergy and Critical Care Medicine, call toll-free 866/CCF-LUNG (223-5864).

For additional information, please visit our Web site at: www.clevelandclinic.org/pulmonary
DEPARTMENT OF Pulmonary and Critical Care Medicine

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Muzaffar Ahmad, M.D.
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Raed A. Dweik, M.D.
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Specialty Interests: asthma, pulmonary hypertension, chronic obstructive pulmonary disease, critical care, bronchoscopy, nitric oxide in lung physiology and disease, exhaled markers in lung disease

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Specialty Interests: asthma, pulmonary hypertension, lung cancer

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Specialty Interests: asthma, pulmonary function tests, pleural disease, bronchoscopic techniques, critical care, pulmonary alveolar proteinosis

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James K. Stoller, M.D.
Head, Section of Respiratory Therapy
Joint Appointment with Medical Division Office - Vice Chairman
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Specialty Interests: bronchoscopy, clinical epidemiology, alpha-1 antitrypsin deficiency, respiratory therapy

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Joint Appointment with Cell Biology, Immunology, Transplant Center
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Specialty Interests: pulmonary defense mechanisms, asthma, adult respiratory distress syndrome, cancer, immunology and chronic infections

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Specialty Interests: asthma, allergic disorders, sinusitis, urticaria, angioedema, latex allergy, aspirin sensitivity
Thanks to a $2 million grant from the U.S. Department of Health and Human Services, The Cleveland Clinic has opened a Sarcoidosis Center of Excellence.

The federal grant will be used to fund an outpatient treatment center for sarcoidosis, develop educational materials to raise awareness, and implement a community outreach program. In addition, the Clinic will use the grant to conduct research on the causes of the disease, the nature of inflammation and the local disease burden.

“We are excited by the opportunity to raise the awareness and understanding of sarcoidosis, the cause of which is currently unknown,” says Mani Kavuru, M.D., a Cleveland Clinic pulmonologist. “Support for our plans has been outstanding, both locally and with the federal grant from the Health and Human Services’ Office of Minority Health. Ultimately, we hope to better understand the disease, treat it more effectively and even prevent it.”

In the United States, African-Americans are eight times more likely than Caucasians to be affected by pulmonary sarcoidosis. Although the disease often resolves on its own, the American Lung Association estimates that 20 percent to 30 percent of patients suffer permanent lung damage as a result of the disease. A small percentage of patients develop chronic sarcoidosis. Currently, there is no prevention or cure.

Opened in February 2003, the Cleveland Clinic’s Sarcoidosis Center of Excellence serves the Northeast Ohio community and is the only one of its kind in the region.

For more information, call 216/445-8747 or 866/783-3679.