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NEW PULMONARY HYPERTENSION GUIDELINE CHALLENGES USE OF COMMON MEDICATION

Genetic Testing Recommended in Patients With Family History of Pulmonary Hypertension

(NORTHBROOK, IL, July 12, 2004) – A new evidence-based guideline for pulmonary arterial hypertension (PAH) cautions the use of calcium channel blockers, a commonly used treatment for high blood pressure, in unstable patients due to the potentially fatal side effects associated with the medication. PAH is a life-threatening condition that occurs when the arteries that supply blood to the lungs become constricted, limiting the blood flow to the lungs and, ultimately, causing high blood pressure to develop within the lung arteries.

The American College of Chest Physicians (ACCP) *Diagnosis and Management of Pulmonary Arterial Hypertension: ACCP Evidence-Based Clinical Practice Guideline* provides recommendations for diagnosing and treating PAH. Published in the July issue of *CHEST*, the peer-reviewed journal of the ACCP, the guideline was developed by a multidisciplinary panel of experts from five medical specialties and is endorsed by the American College of Cardiology Foundation, American College of Rheumatology, American Heart Association, and the Pulmonary Hypertension Association. Panel members recommend against the empiric use of calcium channel blockers or their use in patients who do not respond to acute pulmonary vasodilator testing, citing an increased risk of adverse and potentially fatal events related to the use of the medication. Due to the severity of the disease, the panel also advises genetic testing for patients with a family history of PAH and advance screening for patients with certain chronic diseases who are predisposed to PAH.

“Calcium channel blockers are regularly used to treat high blood pressure because they limit calcium entry into the cells and dilate the constricted systemic blood vessels, thereby lowering blood pressure. This rationale is frequently applied to their use in PAH; however, when they are used in patients with PAH whose narrowed pulmonary arteries are not caused by dynamic vessel constriction, the side effects can be fatal,” said Panel Chair Lewis J. Rubin, MD, FCCP, University of California San Diego School of Medicine, La Jolla, CA. “When left untreated, PAH can cause serious health problems, such as difficulty breathing, blood clots, and fluid retention due to right-sided heart failure. Moreover, patients in specific populations, such as women who are pregnant and patients with respiratory disease, are at greater risk of developing severe complications as a result of PAH. Genetic testing can help to identify patients who are most at risk for developing PAH, allowing clinicians to closely monitor patients and begin treatment at the first sign of the disease.”

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The evidence-based guideline is based on a structured review of all available guidelines and published research related to PAH. The guideline provides specific recommendations for screening, early detection and diagnosis, medical therapies, surgical interventions, sleep-disordered breathing, and prognosis of PAH. Recommendations were graded in regard to the quality of evidence available or expert opinion and the benefit of the diagnostic or therapeutic procedure for the patient population. Aside from genetic testing, the guideline recommends that patients with unexplained PAH undergo testing for connective tissue disease and HIV infection, conditions that may predispose patients to PAH. Patients who are evaluated for PAH also are advised to undergo assessment for sleep-disordered breathing, a potential independent risk factor or complicating factor for PAH. In addition, the guideline supports the continued use of right-heart catheterization to confirm the presence of PAH and establish the severity of the disease. In regard to medical therapies, the guideline recommends that women with PAH avoid becoming pregnant due to the high maternal and fetal mortality rates associated with the disease.

“There are many misconceptions about PAH, since it is relatively uncommon, leading to inappropriate reliance on some tests and treatments. In addition, symptoms of PAH are similar to other respiratory diseases, such as asthma, and can remain subtle for months or even years. These issues have made diagnosing and managing PAH more difficult and the need for early and accurate testing crucial,” said Dr. Rubin. “The extensive published research on PAH over the past few years affords the development of guidelines that are based on data and experience for a very serious disease. When implemented, the guidelines can lead to earlier and more accurate diagnosis of PAH and more appropriate application of therapies now available.”

Although the true incidence of PAH is unknown, it is estimated that over 100,000 people in the United States suffer from PAH and several thousand new cases are diagnosed each year. Pulmonary hypertension can develop in patients of all ages and ethnic groups, and both genders; however, women ages 20 to 40 have the highest incidence of PAH.

“Evidence-based medicine provides a systematic way of practicing medicine based on a comprehensive review of clinical research findings,” said Richard S. Irwin, MD, FCCP, President of the American College of Chest Physicians. “The new evidence-based guideline for pulmonary hypertension combines clinical expertise with extensive external evidence that will enable health-care professionals to practice more effective patient-focused care.”

The development of *Diagnosis and Management of Pulmonary Arterial Hypertension: ACCP Evidence-Based Clinical Practice Guideline* was supported by unrestricted educational grants from Actelion Pharmaceuticals US, Inc., Encysive Pharmaceuticals, and GlaxoSmithKline. To order a copy of the guidelines or for more information, contact the ACCP at 800-343-ACCP (2227) or visit the ACCP Web site at www.chestnet.org.

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