# First guidelines for patients with pulmonary hypertension in sickle cell disease

March 18 2014

Boston Medical Center (BMC) and Boston University School of Medicine (BUSM) physicians have helped create the first set of clinical guidelines for treating patients with pulmonary hypertension in sickle cell disease. Elizabeth Klings, MD, director of the pulmonary hypertension inpatient and education program at BMC and associate professor of medicine at BUSM, spearheaded the development of these guidelines, which are published in the *American Journal of Respiratory and Critical Care Medicine*.

Several studies conducted in the past decade have demonstrated that cardiopulmonary complications, including pulmonary hypertension, are primary risk factors for death in patients with sickle cell disease (SCD). Pulmonary hypertension (PH) affects between six and 11 percent of adults with SCD and is an independent risk factor for death in these patients. This complication is often under-recognized and many patients are not diagnosed early in the course of their disease.

A group of 24 national physician leaders in pediatric and adult hematology, pulmonology and cardiology convened to develop guidelines specific to these patients. Funded by the American Thoracic Society and endorsed by the Pulmonary Hypertension Association and the American College of Chest Physicians, these guidelines represent the most comprehensive pulmonary recommendations thus far.

"I am proud to have collaborated with my colleagues across disciplines to create these guidelines, which will help providers recognize the link

between sickle cell disease and pulmonary hypertension and deliver the optimum care to these patients," said Klings.

Some of the guidelines include screening SCD adults for pulmonary hypertension even if they are asymptomatic every one to three years. All SCD patients with symptoms suggestive of PH, such as exertional shortness of breath and chest pain, should undergo a full PH workup. These patients are often best managed in a specialty center with expertise in the management of PH and SCD. The guidelines also recommend intensifying SCD therapy for all patients with pulmonary hypertension or an elevated pulmonary artery systolic pressure by echocardiography as these patients are also at an increased risk for death. The management of pulmonary hypertension in these patients is dictated by the hemodynamic numbers obtained by a catheterization of the pulmonary arteries. Patients with symptomatic PH should be considered for treatment of their condition.

Provided by Boston University Medical Center