

ATS releases guidelines on the management of pulmonary hypertension of sickle cell disease

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The American Thoracic Society has developed clinical practice guidelines to help clinicians identify and manage patients with sickle cell disease who are at increased risk for mortality from pulmonary hypertension.

"With the development of new treatments, many [patients](#) with [sickle cell disease](#) are now surviving long enough to develop pulmonary hypertension, with an estimated prevalence of 6 to 11 percent," said Elizabeth S. Klings, MD, associate professor of medicine at the Boston University School of Medicine and chair of the committee that produced the guidelines. "Although pulmonary hypertension and elevated tricuspid jet velocity (TRV, an indicator of pulmonary hypertension measured by echocardiography) are both associated with an increased [mortality risk](#), there is currently no standardized approach for identifying and managing these patients."

The guidelines appear in the March 15th issue of the American Thoracic Society's *American Journal of Respiratory and Critical Care Medicine*.

Guideline recommendations include:

- Mortality risk can be accurately determined noninvasively by measurement of the TRV with Doppler echocardiography or by measurement of serum N-terminal pro-brain natriuretic peptide

(NT-pro-BNP) levels and can be determined invasively by direct hemodynamic measurements via right heart catheterization (RHC).

- An increased risk for mortality is defined as a TRV ≥ 2.5 m/second, an NT-pro-BNP level ≥ 160 pg/ml, or RHC-confirmed pulmonary hypertension.
- Patients found to have an increased mortality risk should be treated with hydroxyurea. Patients who do not respond to or are not candidates for hydroxyurea treatment can be considered for chronic transfusion therapy.
- In patients with RHC-confirmed pulmonary hypertension, venous thromboembolism, and no additional risk factors for hemorrhage, indefinite anticoagulant therapy rather than a limited duration of therapy should be used.
- Patients with elevated TRV alone or elevated NT-pro-BNP alone should not be treated with targeted [pulmonary arterial hypertension](#) therapies, including prostanoid, endothelin receptor antagonist, and phosphodiesterase-5 inhibitor therapy
- Most patients with RHC-confirmed pulmonary hypertension should not be treated with targeted therapy
- In select patents with RHC-confirmed marked elevation of pulmonary vascular resistance, normal pulmonary capillary wedge pressure, and related symptoms, a trial of either a prostanoid or an endothelin receptor antagonist may be performed.
- Patients with RHC-confirmed marked elevation of pulmonary vascular resistance, normal pulmonary capillary wedge pressure, and related symptoms should not receive phosphodiesterase-5 inhibitor therapy as first-line treatment.

"As our understanding of sickle cell disease develops, so will our ability to detect disease earlier and to tailor treatment approaches. We need to continue our research efforts into this disease and its management to

understand what the optimal treatment regimen for these patients is. Most of our current recommendations are limited by a lack of large-scale clinical trials in this population." said Dr. Klings. "Management of patients with sickle cell disease with an [increased risk](#) for mortality and [pulmonary hypertension](#) will ultimately be a collaborative effort including adult and pediatric pulmonologists, cardiologists, and hematologists."

Provided by American Thoracic Society

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