Abstract

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Pulmonary Hypertension in Hemolytic Disorders: Pulmonary Vascular Disease: The Global Perspective

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Abstract

The inherited hemoglobin disorders sickle cell disease and thalassemia are the most common monogenetic disorders worldwide. Pulmonary hypertension is one of the leading causes of morbidity and mortality in adult patients with sickle cell disease and thalassemia, and hemolytic disorders are potentially among the most common causes of pulmonary hypertension. The pathogenesis of pulmonary hypertension in hemolytic disorders is likely multifactorial, including hemolysis, impaired nitric oxide (NO) bioavailability, chronic hypoxemia, chronic thromboembolic disease, chronic liver disease, and asplenia. In contrast to patients with traditional forms of pulmonary arterial hypertension, patients with hemolytic disorders have a mild-to-moderate degree of elevation in mean pulmonary pressures, with mild elevations in pulmonary vascular resistance. The hemodynamic etiology of pulmonary hypertension in these patients is multifactorial and includes pulmonary arterial hypertension, pulmonary venous hypertension, and pulmonary hypertension secondary to a hyperdynamic state. Currently, there are limited data on the effects of any specific treatment modality for pulmonary hypertension in patients with hemolytic disorders. It is likely that maximization of treatment of the primary hemoglobinopathy in all patients and treatment with selective pulmonary vasodilators and antiproliferative agents in patients with pulmonary arterial hypertension would be beneficial. However, there is still a major need for large multinational trials of novel therapies for this patient population.

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Biography

Dr. Machado's laboratory focuses on the study of pulmonary hypertension and pulmonary complications of sickle cell disease. His research has contributed to the discovery of pulmonary hypertension as a major risk factor for death in adult patients with sickle cell disease. Using human oligonucleotide microarray platforms, genetics and bioinformatics tools, he currently seeks to develop novel diagnostic, prognostic and therapeutic biomarkers for patients with complex disorders such as pulmonary hypertension. His efforts have led to the identification of novel genetic modifiers and biochemical and molecular biomarkers in sickle cell disease and pulmonary hypertension. His laboratory also utilizes animal models of sickle cell disease and pulmonary hypertension to investigate the role of the pulmonary vascular endothelium in regulating vasomotor tone, permeability, vascular proliferation and apoptosis. By combining these approaches, Dr. Machado seeks to gain novel insights into the pathogenesis and treatment of these complex lung disorders, which would hopefully lead to an improvement in the morbidity and mortality associated with sickle cell disease and pulmonary hypertension.