**1029**

**Multifactorial Pulmonary Hypertension Secondary To Sickle Cell Disease**

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**Purpose:** To understand that there are five groups of pulmonary hypertension (PH) based on different underlying pathophysiologies according to the World Health Organization, and that sickle cell anemia can lead to Group 5 PH.

**Background:** Pulmonary hypertension (PH), defined as elevated pulmonary artery pressures, has different etiologies and is classified into five groups according to the World Health Organization. Sickle cell anemia, via nitric oxide depletion, endothelial dysfunction, inflammation, and increased vascular stiffness can result in Group 5 PH.

**Methods:** A 29-year-old African-American man with history of sickle cell anemia presented with chest, abdominal and bilateral ankle pain. Chest X-ray was normal. An EKG was consistent with left-ventricular (LV) hypertrophy. A trans-thoracic echocardiogram demonstrated LVEF of 58%, elevated right ventricular systolic pressure (RVSP) of 68 mmHg with tricuspid valve regurgitant velocity of 3.80 m/s. Labs showed normocytic anemia, hemoglobin (Hb) 5.8 g/dL, with his baseline Hb between 6 and 7 g/dL. Haptoglobin was <14 mg/dL (normal 30-200), LDH was 349 U/L (normal 122-222). The received intravenous hydration, oxygen, and was continued on his hydroxyurea therapy. The patient was discharged on hospital day three when his pain improved and his hemoglobin stabilized between 6-7 g/dL.

**Results:** Pulmonary hypertension (PH), as defined by elevated pulmonary artery pressures, has many different etiologies. The World Health Organization has classified PH into five different groups. Group 1 PH refers to pulmonary arterial hypertension. Group 2 PH refers to left sided heart disease resulting in elevated left atrial pressures and subsequent compensatory increase in right ventricular systolic pressures. Group 3 PH refers to lung disease resulting in elevated pulmonary artery pressures. Group 4 PH refers to chronic thromboembolic occlusion and PH. Group 5 PH refers to multifactorial PH that includes sickle cell disease. Hemolytic anemia leads to nitric oxide depletion and endothelial dysfunction, inflammation, and increased vascular stiffness found in PH. PH associated with hemoglobinopathies portends a poor prognosis.

**Conclusion:** A possible pulmonary complication of sickle cell disease is Group 5 PH, in which hemolysis leads to pulmonary vasculature dysfunction and subsequent PH.