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Sickle cell cardiomyopathy: Past, present, and future

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Softhe cell disease (SCD) is an inherited chronic hemolytic anemia whose clinical manifestations arise from the tendency of the hemoglobin to polymerize and deform red blood cells into the characteristic sickle shape due to a single nucleotide change in the beta-clobin. This leads to vascular occlusion of small and large vessels which leads to chronic damage to several organs including the brain, lungs, kidneys, liver, and spleen. Chronic sickle cell anemia leads to several cardiovascular manifestations. These include high cardiac output, cardiomegaly, and progressive iron overload in patients after multiple blood transfusions leading to secondary hemochromatosis. These patients develop right and left sided systolic and diastolic dysfunction. Pulmonary hypertension is another complication of chronic sickle cell anemia which is a poor prognostic factor and in turn may lead to worsening right heart failure. Treatment strategies include limiting blood transfusions, iron chelation, and exchange transfusions. We present data from our center depicting the proportion of systolic dysfunction in our patients with sickle cell disease. We also present a case series of patients from our center with improvement in secondary hemochromatosis after iron chelation. Work is underway to assess the benefits of cardiac medications in patients with sickle cell cardiomyopathy, novel agents that target the biology of vasculopathy to prevent the cardiac complications of sickle cell disease, as well as gene therapy in the near futures.

Biography

Mahazarin Ginwalla, MD, MS is an Assistant Professor and Medical Director of Mechanical Circulatory Support at the University Hospitals Case Medical Center in Cleveland, Ohio. She received her medical degree from Lokmanya Tilak Medical College in India. She completed her Masters in Medical Microbiology and Immunology at the University of Wisconsin-Madison, and subsequently completed her Internal Medicine residency at the Medical College of Wisconsin. She trained in cardiology at Case Western Reserve University/ Metrohealth Medical Center, and in Advanced Heart Failure and Cardiac Transplantation at Stanford University Medical Center. She has been actively involved in clinical work, education, and research in the areas of heart failure especially sickle cell cardiomyopathy, mechanical circulatory support, and cardiac transplantation.

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