TITLE: Management of Cerebral Vasculopathy in Pediatric Sickle Cell Disease

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RESEARCH PROJECT DESCRIPTION

Children with sickle cell disease (SCD) are at high risks for both stroke and cerebral vasculopathy. Cerebral vasculopathies, including Moyamoya Disease and intracerebral aneurysms can lead to similar SCD morbidities such as ischemic stroke and intracerebral hemorrhage. For those SCD patients already suffering cerebral vasculopathy, their risk for stroke becomes greater. Previous studies indicate that both decreases in hemoglobin S and cerebral revascularization procedures may reduce the risk of stroke in pediatric SCD.

The purpose of this longitudinal study is to determine the optimal management and surgical interventions for patients diagnosed with both cerebral vasculopathy and SCD. In order to do so, we will need to (1) examine the rate of cerebral vasculopathy among pediatric SCD patients identified to be high risk for stroke and monitor the development of cerebral vasculopathy in SCD; and (2) to determine the effectiveness of cerebral revascularization procedures in pediatric patients with both sickle cell disease and cerebral vasculopathy.

The role of the medical student for this study would include assisting with data collection, uploading data into the registry, and potentially assist with manuscript development.

Kassim AA and DeBaun MR. Sickle cell disease, vasculopathy and therapeutics. Annu Rev Med, 64:26.1-26.16, 2013.

Smith ER, MCClain CD, Heeney M and Scott RM. Pial synangiosis in patients with moyamoya syndrome and sickle anemia: perioperative management and surgical outcome. Neurosurg Focus 26 (4):10, 2009