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114.SICKLE CELL DISEASE, SICKLE CELL TRAIT AND OTHER HEMOGLOBINOPATHIES, EXCLUDING THALASSEMIAS: CLINICAL AND EPIDEMIOLOGICAL

Microemboli on Transcranial Doppler in Patients with Sickle Cell Disease: A Single Center Case Series

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Background: Stroke remains a major cause of morbidity in patients with sickle cell disease (SCD). Current guidelines recommend that patients with Hemoglobin SS and Hemoglobin S beta zero thalassemia have annual transcranial doppler (TCD) screening and that patients with abnormal velocities should have regular blood transfusions to maintain a maximum HbS level <30% and maintain a hemoglobin level >9 g/dL for at least one year to decrease risk of stroke (DeBaun et al., 2020). While research has shown that patients with velocities >200cm/sec are at increased risk of cerebral infarct, there is no literature to discuss the follow up and management of patients with microemboli on TCD.
In the past microemoli were unable to be detected, however TCD is now able to detect such findings (Wan et al., 2022). Studies have been completed on patients with ischemic stroke without SCD and have shown that the presence of microemboli on TCD is associated with a higher rate of recurrence (Das et al., 2020). The presence of emboli has also been shown to recognize patients with asymptomatic carotid stenosis and increased risk for stroke (Markus et al., 2010). Although noted in the general population, the significance of such a finding on an annual TCD screening in patients with sickle cell disease is unknown.
Methods: At the Medical University of South Carolina, we performed a retrospective review of all patients with SCD who have had TCDs from 2017 to February 2023 and were found to have microemboli to evaluate management and outcome.
Results: We have had 5 patients with an incidental finding of microemboli on their annual TCD since 2017. Three were male with median age of 3 (range 2 to 15 years). Four individuals are HbSS genotype. Mean hemoglobin at the time of TCD was 8.26 ± 1.4 g/dL. TCD velocity was normal in four patients at the time of the TCD, while the remaining individual had a low conditional TCD. Two have a previous history of conditional or abnormal TCDs. The location of the microemboli was variable among patients (Table 1).
At the time of microemboli findings on TCD, discussions were held with family regarding options for treatment including chronic partial exchange transfusions and optimization of other disease modifying therapies. Management of all patients included MRI at the time of microemboli, which was normal in only two patients. Three patients were started on chronic manual exchange transfusions for 10 months to 3 years. One out of the 3 continued home HU while receiving transfusions while the remainder were transitioned to HU once chronic transfusions were complete. One patient was not started on chronic transfusions but monitored closely with TCDs due to family choice. On follow up, 4 have had repeat TCDs and all have normalized without any subsequent episodes of microemboli. One patient was found to have silent infarct on MRI at time of TCD and 1 has had follow up MRI 4 years after original TCD with a new silent infarct. Of note, this patient had inconsistent follow up with chronic transfusions after initial abnormal TCD. Importantly, none of our 5 patients had an acute symptomatic ischemic event since the microemboli were noted. Overall, among these patients, 2 have had concern for silent infarct and 3 of 5 started on chronic transfusions (Table 1).
Conclusion: This is the first report within the literature of microemboli on annual TCD screening in patients with SCD. As this is a new phenomenon, the significance within the disease course is not known for those with sickle cell disease. Identifying patients with microemboli has been used to help navigate the need for early intervention to prevent stroke. Given that individuals with SCD are already at an increased risk for cerebrovascular events, these findings of microemboli could indicate further risk in these individuals as well and deserve further investigation. Further research is warranted as the significance of such a finding is unknown and further understanding could impact the standard of care of this patient population.

Disclosures Johnson: *Pfizer/GBT:* Membership on an entity's Board of Directors or advisory committees.

Patient	Year	Genotype	Age (years)	Sex	Hb (g/dL)	TCD velocity at time	TCD normalized	Location of microemboli	MRI	Intervention	Silent infarct	Acute ischemic infarct

Figure 1

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