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

Poor School Performance As an Indicator of Severity of Sickle Cell Disease

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Purpose: Patients with sickle cell anemia can have long term neurocognitive deficits throughout their lifespan. The purpose of this study is to conduct neuropsychiatric evaluations and brain imaging to determine if individuals who have higher risk of falling behind in education can be identified, emphasize the importance of addressing educational needs of these patients, and identify patients who are at risk of developing further complications.
Methods: IRB approved retrospective study of patients with Hemoglobin SS disease, who underwent Stanford-Binet Intelligence testing and Woodcock Johnson Achievement. All patients who underwent cognitive testing and also received a transcranial doppler ultrasound or magnetic resonance imaging between were included. All patients were referred to neuropsychiatric testing due to poor school performance.
Results: 33 patients were included who underwent neuropsychiatric testing. These students were referred due to poor school performance. Of those, 15 patients had additional imaging studies. All patients were identified with hemoglobin SS disease. All patients were African American. The median age was 9 with an age range of 6 to 16 with a median grade was 4. This sample included 12 females and 3 males. 73% (11 of 15 individuals) were identified with below average full scale intelligence quotient. Of these, 46% (6 of 11) had measurable changes in MRI or transcranial doppler results. 5 of them had measurable changes in transcranial doppler and changes in their MRI, while 1 patient had a normal transcranial doppler, but showed changes in their MRI. On average, the maximum mean velocity was 164 cm/s and was higher than the patients with normal intelligence quotient scores at 148cm/s. The remaining 5 individuals also had below average full scale intelligence quotient without any findings on imaging.
All 11 individuals also had below average scores in academic application, academic fluency, and academic skills. On average these patients were 3 years behind on reading, 2 years behind on math, and 2 years behind writing. Only one of these patients had an individualized education plan with extra assistance for education.
7 of the 11 patients with below average full scale intelligence quotients were on hydroxyurea and 2 of the 4 patients with average scores used hydroxyurea. The average hemoglobin level in the group that performed below average was 9.5 and 8.9 in the group that performed average. While, all patients who had average scores on the stanford-binet test did not have any abnormal transcranial dopplers and they all had delays in math, reading, and writing.
Conclusion:
Below average intelligence and delays in academic performance may be an indicator of patients at risk of developing further complications such as stroke.The full extent to this relationship is unclear. Further research is needed to determine the relationship between intelligence, academic performance and changes in imaging. Earlier detection of these patients can prevent further complications, develop individual treatment plans, and target closer monitoring. A multidisciplinary approach to treating patients with sickle cell disease should include neuropsychiatric evaluations, an assessment of academic achievement, and the opportunity for additional support in school to fully address the needs of this population.

Disclosures Crary: *Takeda:* Membership on an entity's Board of Directors or advisory committees; *Bayer:* Membership on an entity's Board of Directors or advisory committees; *Novartis:* Membership on an entity's Board of Directors or advisory committees, Other: DSMB consulting; *GBT:* Membership on an entity's Board of Directors or advisory committees.

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