



DATE: March 7, 2022 **COMMITTEE:** House Health and Government Operations
BILL NO: House Bill 1185
BILL TITLE: Public Health – Sickle Cell Disease – Referral for Transcranial Doppler Ultrasound
POSITION: Support

Kennedy Krieger Institute supports House Bill 1185 - Public Health – Sickle Cell Disease – Referral for Transcranial Doppler Ultrasound

Bill Summary:

House Bill 1185 requires physician or advance practice nurse to refer patients who are minors that have sickle cell disease to a hematologist for a transcranial Doppler ultrasound.

Background:

Kennedy Krieger Institute is dedicated to improving the lives of children and young adults with developmental, behavioral, cognitive and physical challenges. Kennedy Krieger’s services include inpatient, outpatient, school-based and community-based programs. Over 25,000 individuals receive services annually at Kennedy Krieger.

The Kennedy Krieger Institute Sickle Cell Neurodevelopmental Clinic provides neurological, developmental, neuropsychological, and behavioral psychology services in a comprehensive, multi-disciplinary outpatient clinic. We also see the siblings of children who have neurodevelopmental disorders, many of whom have sickle cell trait, both in the Sickle Cell Neurodevelopmental Clinic and other Kennedy Krieger outpatient clinics.

Rationale:

Children with sickle cell disease have high rates of neurological and neurodevelopmental complications.^{1,2} Stroke, including silent stroke, is quite frequent in these patients, with devastating consequences requiring lifelong care and medical follow up. Transcranial Doppler ultrasound testing has been shown to be predictive of risk for future stroke, allowing high-risk patients to receive prophylactic blood transfusions to decrease stroke risk.^{3,4} These specialty ultrasounds must be completed by a well-trained technician/clinician with results communicated to a hematologist to determine appropriate management and follow up needs. All patients with certain subtypes of sickle cell disease should be receiving transcranial Doppler ultrasound screenings per the most recent sickle cell disease guidelines.⁵ Improvement in referral rates for transcranial Doppler ultrasounds could decrease significantly the rate of brain injury in these patients, impacting future disability, quality of life, and life expectancy.

Kennedy Krieger Institute requests a favorable report on House Bill 1185.

1. Ohene-Frempong K, Weiner SJ, Sleeper LA, et al. Cerebrovascular Accidents in Sickle Cell Disease: Rates and Risk Factors. *Blood*. January 01 1998;91(1):288.
2. Lance EI, Cannon AD, Shapiro BK, Lee LC, Johnston MV, Casella JF. Co-Occurrence of Neurodevelopmental Disorders in Pediatric Sickle Cell Disease. *J Dev Behav Pediatr*. Aug 01 2021;42(6):463-471. doi:10.1097/DBP.0000000000000914
3. Adams RJ, McKie VC, Brambilla D, et al. Stroke prevention trial in sickle cell anemia. *Controlled clinical trials*. Feb 1998;19(1):110. doi:S0197245697000998 [pii]
4. Adams R, McKie V, Nichols F, et al. The Use of Transcranial Ultrasonography to Predict Stroke in Sickle Cell Disease. *N Engl J Med*. 02/27; 2011/10 1992;326(9):605. doi:10.1056/nejm199202273260905
5. DeBaun MR, Jordan LC, King AA, et al. American Society of Hematology 2020 guidelines for sickle cell disease: prevention, diagnosis, and treatment of cerebrovascular disease in children and adults. *Blood advances*. Apr 28 2020;4(8):1554. doi:10.1182/bloodadvances.2019001142 [doi]