

BURDEN AND RISK OF NEUROLOGICAL AND COGNITIVE IMPAIRMENT IN PEDIATRIC SICKLE CELL ANEMIA IN UGANDA (BRAIN SAFE): INTERIM OVERALL RESULTS

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Background: Sickle Cell Anemia (SCA) is common in sub-Saharan Africa (SSA). In Uganda, 20,000 children are born with SCA annually. Sickle brain vasculopathy causes both overt strokes and “silent infarcts”, affecting neurological and cognitive function. Our objectives are to determine the age-related spectrum and burden of brain injury in Ugandan children with SCA.

Methods: In first year we screened 248 children with SCA, aged 1-12 years, attending the Mulago Hospital SCD clinic in Kampala. We excluded age >12, acute illness, Hb <6.0gm/dl, recent transfusion. A stroke history and examination were performed using the pediatric NIH Stroke scale. Psychometric testing using age-appropriate KABC-II, TOVA, BRIEF and Mullen testing and Transcranial doppler ultrasounds (TCDs) were performed. Brain magnetic resonance (MR) imaging was performed on a subset with a history of stroke, abnormal neurological exam or cognitive testing.

Results: We enrolled 233 (mean age 5.62 years), 7 had a history of stroke, and an abnormal neurological exam, 5 had a history of stroke but a normal neurological exam, 6 had abnormal neurological findings but no history of stroke (total abnormal 18/233, 7.7%). To date, cognitive test data results have been performed on 80 children; 30 (37.5%) were impaired. A total of 224 TCDs have been performed. 190 were normal, 4 abnormal, and 30 conditional. A subset of 29 subjects were selected with either clinical and/or historical stroke pathology (N= 25, 86%) and have undergone MR imaging. Mean age was 7.1 years (range 3-12); male: 55%. Of 26 with clinical radiological reads, 16/26 (61.5%) were abnormal. Of 12 subjects, 2 had no vasculopathy; 10 were abnormal: 6 infarcts and arterial stenoses, 3 infarcts, 1 bilateral stenoses.

Conclusion: Preliminary results demonstrate brain vasculopathy in SCA, a high prevalence of abnormal history or physical findings, abnormal psychometrics, abnormal cerebral blood flow and/or brain MR imaging.