Neuropsychological dysfunction in children with sickle cell disease : Diagnostic impact of TCD, MR and SPECT imaging

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Sickle cell disease (SCD) is a genetic blood disorder caused by abnormal hemoglobin that damages and deforms red blood cells (RBCs) and can lead to anemia and obstruction of blood vessels which can cause cerebrovascular stroke to those patients with the subsequent deterioration of neurocognitive functions and academic underachievement.

Moreover, recent evidence suggests that patients with hemoglobin SS sickle cell disease may have cognitive dysfunction, even if they are free of infarction. The aim of this study was to evaluate the neuropsychological functions of children with sickle cell disease and its correlation to neuroimaging abnormalities using MRI, SPECT and TCD procedures.

Twenty-three patients with SCD and nine healthy matched controls were recruited and assessed using Wechsler intelligence scale for children III(WISC III), Wisconsin card sorting test (WCST) and wide range achievement test- 4(WRAT4), all test scores were then correlated to MRI, MRA, SPECT,TCD findings and to disease severity.

There was statistical significant decline in scores of WISC, WRAT4, and WCST in patients group compared to controls. A decline in cognitive functions in children with sickle cell disease was also statistically correlated to neuroimaging abnormalities and to disease severity. The findings supported the need for periodic neuropsychological evaluation in patients with sickle cell disease in order to identify the patterns of higher cortical dysfunction and assist in the development of appropriate rehabilitation and special education programs for patients with SCD even in the absence of CVA.