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The Language and Working Memory Abilities of Children with Sickle Cell Disease with and without Silent Cerebral Infarct: A Preliminary Study

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Introduction

Sickle cell disease (SCD) is a recessive inherited blood disorder that can cause a sequelae of central nervous system complications. Recent studies have suggested that 22\% of all school-aged children with SCD (hemoglobin SS) will experience a silent cerebral infarct (SCI) in the absence of any overt neurological or physical symptoms before their 14\textsuperscript{th} birthday (Adams, 2001; Armstrong, et al., 1996; DeBaun, et al., 1998). Approximately 75\% of these children will demonstrate neuropsychological deficits that can have a devastating impact on their academic achievement (Steen, 2003; Schatz, 2004; Nettles, 1994).

This investigation sought to (1) determine whether children with SCD with and without SCI would differ on tasks that evaluated language and working memory; (2) determine the relationship of neurocognitive and biological markers to the language outcomes of children with SCD with and without SCI.

Methods

All the participants with SCD were recruited from the Silent Transfusion Trial (SITT) at the National Children’s Medical Center (CNMC) in Washington, DC. Thirty-two participants with diagnosed SCD hemoglobin SS or sickle $\beta^0$ Thalassemia (hemoglobin S$\beta^0$) and no history of stroke were recruited for this study. All participants underwent a brain magnetic resonance imaging (MRI) screening and a transcranial doppler screening (TCD). Participants were divided into two groups, Group 1: Children with SCD and no evidence of SCI (n=16); and Group 2: Children with SCD and evidence of SCI, a negative neurological examination and TCD velocities of < 200cm/sec (n=16). Hematocrit levels (Hct), the number of hospitalizations (Hosp P) and pain episodes over the past year were recorded. Controls were recruited from siblings of participants and the community.

All participants were administered The Clinical Evaluation of Language Fundamentals, 4\textsuperscript{th} edition (CELF-4, Semel, Wiig & Secord, 2001) to measure language function and verbal working memory and the Weschler Intelligence Scales (WAIS, Weschler, 1999) to measure intellectual abilities.
Results

The results revealed that children with SCD without SCI had significantly lower language scores than the control group and children with SCD and SCI. On measures of verbal working memory (VWM) there were no significant differences between the groups of participants.

A preliminary investigation revealed verbal working memory (VWM) ($r=.61$), IQ ($r=.61$) and Hosp P ($r=.57$) were positively related to language performance.

Conclusion

The study suggests that factors (frequency of hospitalizations, verbal working memory) other than SCI in children with SCD may have the greatest negative effect on language performance and academic achievement.

References


