Hydroxyurea use is associated with executive functioning and nonverbal skills in young children with sickle cell disease (SCD)

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Abstract

Cognitive impairment in children with sickle cell disease (SCD) is thought to reflect the complex pathophysiology of the disease. Hydroxyurea (HU) is used in children with SCD to increase fetal hemoglobin (HF), contributing to a decrease in physical symptoms and to potential protection against cerebral microvasculopathy. There has been minimal investigation into the association between HU use and cognition in this population. This study examined the relationship between HU status and cognition in young children with SCD. Neuropsychological data were collected in a prospective study from 38 children with SCD HbSS or HbS/?0 thalassaemia ages 4 to 11 years with no history of overt stroke or chronic transfusion. Controlling for hemoglobin, children on HU performed significantly better than children who were not on HU on measures of attention/executive functioning and nonverbal skills. Performance on verbal measures was similar between groups. Though not statistically significant, duration of HU use was positively associated with better performance in all domains. These results provide preliminary evidence that treatment with HU may not only reduce physical symptoms, but may also be protective against cognitive dysfunction in young children with SCD, particularly in regard to attention/executive functioning and nonverbal skills. Replication with larger samples and longitudinal studies are warranted.

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