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## Original article

# Comprehensive neuropsychological evaluation of children and adolescents with sickle cell anemia: a hospital-based sample

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ABSTRACT

*Background:* Individuals with sickle cell anemia may suffer symptomatic or silent cerebral infarcts leading to neurocognitive complications. This study investigated the cognitive and intellectual performance of children and adolescents with sickle cell anemia.

*Methods:* The socioeconomic status, clinical aspects and behavioral profile of 15 young individuals with sickle cell anemia were evaluated. The Wechsler Intelligence Scale for Children, the Developmental Neuropsychological Assessment Test, and the Child Behavior Checklist were applied.

*Results:* Participants with a history of stroke had lower intelligence quotient (IQ) scores. Alterations were found in attention and executive functioning, language, verbal and visual memory, visuospatial processing and sensorimotor skills. These alterations were found both in the children and adolescents who had had a cerebral infarction and in those who apparently had not. In the majority of cases, there were learning difficulties, a history of repeating school years and a need for specialist educational support. The most common additional diagnoses in accordance with the Diagnostic and Statistical Manual of Mental Disorders IV were depressive disorder, anxiety disorder and somatic disorder, as well as conditions associated with physical and psychosocial repercussions of sickle cell anemia.

*Conclusion:* As sickle cell anemia is considered a progressive cerebral vasculopathy, it is a potential risk factor for neurocognitive and psychosocial development. Therefore, periodic neuropsychological and behavioral evaluations of children and adolescents with sickle cell anemia may represent a useful measure to reduce long-term biopsychosocial repercussions.

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## Introduction

Sickle cell disease (SCD) comprises a group of inherited blood disorders resulting from a genetic alteration in hemoglobin. It is the most common monogenic hereditary disease in Brazil.[1] In the state of Bahia, the number of individuals with the sickle cell trait is estimated at 5.5% of the general population and 6.3% of the population of African descent. The incidence of sickle cell anemia (SCA) in Bahia is the highest in Brazil: 1 in 650 live births.[2]

Ohene-Frempong et al. reported that 11% of individuals with SCA develop symptomatic cerebral infarcts prior to completing 20 years of age. The sequelae of these events affect different aspects of the patient's life. Overall, 11–35% of children and adolescents with SCD present with silent strokes identified only by imaging exams and with no observable clinical symptoms.[3]

The neurocognitive complications and their consequences are progressive and originate from a state of cerebral vasculopathy, negatively affecting child development and quality of life.[4]

The principal objective of this study was to evaluate cognitive function and the behavioral profile of children and adolescents with sickle cell anemia.

## Methods

The Internal Review Board of the *Plataforma Brasil* of the Professor Edgard Santos Teaching Hospital, Universidade Federal da Bahia approved the protocol of this descriptive, observational, hospital-based study (#314.636).

Fifteen children and adolescents with ages ranging from 6 to 16 years and SCA diagnosed by hemoglobin electrophoresis participated in this study. Children with visual or auditory deficits, epilepsy, a history of brain trauma, meningitis or intoxications, a history of gestational or neonatal anoxic brain injury, prematurity, concomitant genetic syndromes or any other condition that would prevent the psychometric tests from being conducted were excluded from the study.

The children's parents or guardians were interviewed at admission. The child's birth registration card was reviewed to obtain data related to pregnancy and neonatal conditions. The school report was used to evaluate neuropsychomotor development and educational history, data on the clinical characteristics of SCD were obtained and the socioeconomic criteria of the Associação Brasileira de Empresas de Pesquisa (ABEP) were applied.[5]

A short form of the Wechsler Intelligence Scale for Children (Third Edition – WISC-III) was used to estimate the intelligence quotient (IQ). This instrument is used as a screening tool to evaluate the intellectual performance of Brazilian children (Table 1).[6,7]

Fifteen subtests of the developmental neuropsychological assessment test (second edition – NEPSY-II)[8] were selected to investigate cognitive function. This tool takes a broad and flexible approach to neuropsychological evaluation. Percentiles were used to classify performance in this assessment (Table 1).

The Child Behavior Checklist (CBCL), a questionnaire consisting of 113 items, developed to evaluate individuals in the 6–18 age range, was applied to parents or guardians[9] (Table 1).

## Results

Of the fifteen cases evaluated, nine (60%) were male. The mean age of the patients was nine years (range: 6–16 years). Of the 15 patients, 12 (80%) were right-handed, while three (20%) became left-handed after having suffered a left hemisphere stroke. In these cases, the trained left hand was considered the dominant hand.

In nine (60%) cases, the family reported that the child needed specialist educational support due to difficulties at school.

The IQ test was found to be within the normal range for ten (66.6%) participants, while four (26.6%) had a lower than average score, and one (6.6%) scored above average. Of the four (26.6%) participants who scored below average, three (20%) had a borderline score (70–79). The mean IQ score was 88.5.

In the majority of cases, the individual's performance with respect to immediate memory was poor for visual, spatial and verbal content as well as in the subtests of attention and executive functioning, language and sensorimotor skills (Table 2). Overall, nine (60%) participants performed below average in at least one of the NEPSY-II subtests (Table 3).

The behavioral profile and diagnostic criteria based on the Diagnostic and Statistical Manual of Mental Disorders (fourth Edition) are described in Table 4.

**Table 1 – Classification of performance according to the Wechsler Intelligence Scale for Children (Third Edition) intelligence quotient (IQ), Developmental Neuropsychological Assessment Test (second edition) and Child Behavior Checklist.**

| Estimated IQ[a] | | NEPSY-II[b] | | CBCL[c] | | |
|---|---|---|---|---|---|---|
| Index | Classification | Percentile | Classification | T-score | Percentile | Classification |
| ≤79 | Low | ≤25th | Low | ≤64 | ≤85th | Normal |
| 80–119 | Average | 26–75th | Average | 65–69 | 86–96th | Borderline |
| ≥120 | High | 76–99th | High | ≥70 | ≥97th | Clinical |

a WISC-III: Wechsler Intelligence Scale for Children.[7]
b NEPSY-II: a developmental neuropsychological assessment.[8]
c CBCL: Child Behavior Checklist.[9]