***A Pilot Study of Assessing Cognition in Children with Sickle Cell Disease Using a New Software Package the Cogstate Battery***

1. Jessica Thompson
2. Catherine Mkandawire
3. Subarna Chakravorty, MDPhD
4. Michael Laffan, MD

Introduction

17% of children with sickle cell disease (SCD) between the ages of 6 and 16 could have silent infarcts1. Depending on the area of the brain affected silent infarcts can cause problems with attention, coordination, visual-motor speed and executive function. Children with SCD in the UK do not receive routine MRI scans. Subtle defects in cognition can be assessed with neuropsychometric testing which involves multiple tests assessing many areas of cognition. However, testing is limited to those with known neurological deficits due to lack of funding and shortage of specialist staff. There is a need for a robust screening tool for assessment of cognition, which could identify children for further specialist testing.

The Cogstate battery is computer-based program that assesses cognition and has been used in several clinical settings, both adult and paediatric2-3. The Cogstate battery is reported to be culturally neutral and is available in several languages. Additionally the Cogstate battery is free from practice effects and so could be used as an annual assessment tool in order to identify any declines as early as possible. The Cogstate battery has not yet been used to assess cognition in children with SCD.

Research Objectives

The aim of this study was to assess the feasibility of using the Cogstate Battery as a tool for the assessment of cognition in children with SCD. It was hypothesised that the Cogstate battery would be easy to use within this setting and would be acceptable to patients, parents and assessors.

Methods

Eight clinically well children, aged 10-17 with SCD were recruited through St Mary's Hospital paediatric haematology outpatient clinics. The Cogstate software was downloaded onto a Windows laptop computer and an anonymous profile was created for each child before testing. A battery of 6 tests (Table 1) was created aiming to assess a range of cognitive domains within a reasonable amount of time. Every child completed the battery of tests once, which included a short practice before each test. After testing each patient was asked to give an opinion of how they found the tests. Upon completion of the test the patients' results were uploaded to the Cogstate website which generated a test report and a case report form. A mark was given for each test and a score of over 90 represents normal cognition in the area tested, 81-90 represents mild impairment and below 81 represents impairment.

| **Test Name** | **Cognitive Domain Tested** |
| --- | --- |
| Continuous paired associate learning (CPAL) | Paired associate learning |
| Detection (DET) | Psychomotor function |
| Groton maze learning test (ME) | Executive function |
| Groton maze learning test-delayed recall (ME) | Delayed recall |
| Identification (IDN) | Attention |
| One card learning (OCL) | Learning |
| One-back memory (ONBA) | Working memory |

Table 1.

Tests used in the Cogstate battery and corresponding cognitive domains assessed

Results

8 patients completed the battery, taking on average 29 minutes (Table 2). The battery was easy to carry out and although some children reported it as boring, they all finished the tests without distress. The test report generated by the Cogstate website allowed results to be analysed quickly and with ease. An overall score from each test is clearly indicated. The Continuous Paired Associate Learning test was not displayed as part of the test report as there was insufficient normal data to draw conclusions from the results within the age group tested.

| **Patient ID** | **DET (Psychomotor function)** | **IDN (Attention)** | **OCL (Learning)** | **ONBS (Processing speed)** | **ONBA (Working memory)** | **ME (Executive function & delayed recall)** |
| --- | --- | --- | --- | --- | --- | --- |
| 0001 | 87 | 75 | 82 | 78 | 83 | 99 |
| 0002 | 106 | 105 | 94 | 90 | 117 | 99 |
| 0003 | 96 | 101 | 95 | 95 | 117 | 98 |
| 0004 | 76 | 76 | 94 | 81 | 94 | 93 |
| 0005 | 86 | 84 | 98 | 81 | 89 | 89 |
| 0006 | 98 | 104 | 97 | 97 | 94 | 111 |
| 0007 | 94 | 90 | 91 | 83 | 101 | 86 |
| 0008 | 91 | 88 | 108 | 90 | 96 | 95 |

Table 2.

Summary Report of 8 patients tested

Conclusion

The Cogstate battery is a feasible tool for paediatric SCD patients and can be undertaken in a clinic setting. This feasibility study will help design a prospective, comparative study of cognition in children with SCD using the Cogstate battery and conventional neuropsychometric assessment and once validated, would be a useful tool to assess cognition and institute timely educational and medical intervention.