Poorer executive functioning in preschool children with sickle cell anaemia: evidence from the Preschool Executive Task Assessment

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Running title:

Assessment of executive functions in sickle cell anaemia

Document Word Count: 6,572

Background: Sickle cell disease is the most common hereditary disorder in the United Kingdom, with over one in 2,000 infants diagnosed at birth. Sickle cell anaemia (SCA), the most frequently occurring and severe form of sickle cell disease, is associated with a high risk of stroke. However deficits in executive function are commonly reported in older children with SCA even when there is no evidence for stroke. Earlier identification of these deficits would give more scope for intervention, but their extent in preschool children with SCA has not been well-studied, in part due to a lack of age-appropriate standardized assessments. This paper introduces the Preschool Executive Task Assessment (PETA), which assesses executive skills including a child's ability to maintain attention and follow through on tasks, reflecting real-life challenges that a child may encounter at home or at school.

Methods: The PETA was administered in two studies: In Study 1, 166 typically developing three- to five-year-olds were assessed to validate the PETA and in Study 2 performance of 22 children with SCA was compared with that of 24 ethnicity, age, SES, and IQ-matched comparison children. **Results:** Study 1 showed evidence for task reliability and validity and showed improved performance with age. Study 2 observed poorer executive performance in children with SCA particularly in the domains of completion, sequencing and distractibility.

Conclusions: The PETA can be used to robustly measure executive functioning in an ecologically valid way in preschool children, and reveals that executive deficits are present already at preschool-age in children with SCA

Key Words: Executive function, assessment, preschool children, sickle cell disease, neurodevelopmental disorders

Abbreviations: PETA=Preschool Executive Task Assessment; SCA=Sickle Cell Anaemia; EF=Executive Functioning

Introduction

Sickle cell disease is the most common hereditary disorder in the United Kingdom, with approximately one in 2,000 infants diagnosed. It is a genetic blood disorder in which the haemoglobin in red blood cells, which transport oxygen around the body, is abnormal. Sickle cell anaemia (SCA), the most frequently occurring and severe form of sickle cell disease, is associated with a high risk of stroke. Up to 40% of children with SCA experience clinical or more subtle 'silent' stroke before the age of 15 (Armstrong et al., 1996; Bernaudin et al., 2000), and the fronto-parietal regions are most frequently affected(Ohene-Frempong et al., 1998). Even in the absence of stroke, there is evidence of bilateral cortical thinning and reduced white matter integrity resulting from chronic hypoxemia in frontal, parietal and temporal lobes (Baldeweg et al., 2006).

In addition to a reported decline in IQ, specific deficits in executive functioning (EF) are the most frequently reported cognitive deficits in school-age children and adolescents with SCA, even when there is no evidence for neurological morbidity (Berkelhammer et al., 2007; Hijmans et al., 2011; Nabors & Freymuth, 2002; Noll et al., 2001). The executive system is responsible for combining different faculties in order to execute cognitive control in high level processes such as making plans and solving problems(Welsh & Pennington, 1988). The fronto-parietal network, which is vulnerable to damage in SCA, is known to play a role in mediating executive functions. Most studies of SCA have focused on evaluating EF deficits in school-aged or older patients. Sickle cell researchers have highlighted the difficulty in ascertaining the extent of earlier executive deficits due to the lack of age-appropriate standardized tests (Schatz & Roberts, 2007). The available literature for preschoolers and toddlers with SCA without stroke reports lower IQ scores and developmental quotients, and poorer school readiness skills(Glass et al., 2012; Steen et al., 2002; Tarazi, Grant, Ely, & Barakat, 2007; Thompson et al., 2003). In the only study that has specifically looked at EF in children with SCA younger than school age, infants with SCA tested at nine months and again at twelve months (n=14) showed preliminary evidence for a delay in the development of early EF skills on classical 'A not B' and object retrieval tasks (Hogan, Telfer, Kirkham, & de Haan, 2012).

Research suggests that throughout infancy and the preschool years, performance on executive tasks can be predictive of future EF and school readiness(Blair, 2002; Clark, Prior, & Kinsella, 2002; Kraybill & Bell, 2013). Executive skills are susceptible to environmental and disease factors(Hughes & Ensor, 2009; Stuss & Alexander, 2000). However, unlike IQ, executive skills show potential for improvement with intervention (Diamond & Lee, 2011) and are an important protective factor in children who have lower IQs or deficits in other areas(Greenberg, 2006; Johnson, 2012). It is therefore important to identify executive deficits as early as possible. Whether specific executive skills are already established in preschoolers or emerge from a more undifferentiated system with development is still debated. However, several researchers have found evidence for a unitary construct that becomes more differentiated over time(Senn, Espy, & Kaufmann, 2004; Wiebe et al., 2011). Investigating EF in the preschool years is thus important for understanding whether the deficits seen at later ages reflect an early-existing deficit or one that emerges as result of the differentiation within the EF systems that occurs with development.

Measuring executive functioning in the preschool years

Preschool assessments that rely solely on summary scores are considered to be questionable as an accurate indicator of cognitive abilities in this age group (Carlson, 2005). Ecologically valid measures that adopt a micro-analytic approach incorporating quantitative and qualitative scoring could better represent specific skills developing at various rates and enhance the diagnostic utility of tasks in this age range(Pritchard & Woodward, 2011). Ecological validity is the extent to which performance on a cognitive task reflects real-life performance and can be measured by a task's verisimilitude and veridicality(Chaytor & Schmitter-Edgecombe, 2003). Verisimilitude concerns the similarity between task demands and everyday demands, while veridicality is the strength of association between task performance and everyday functioning. An ecologically valid task may be more engaging for a young child and reflect real-life performance to a greater extent, providing more accurate information on the type and amount of support a child requires in everyday life. A task that includes a global measure as well as a microanalysis of performance is particularly useful in light of theoretical models of EF that suggested subcomponents within this system become more differentiated with age. The few existing standardized tasks for this age range do not easily allow this is they tend to focus on specific aspects of EF in isolation.

Ecologically valid measurement of executive functioning

Ecologically valid measures that reflect real-life tasks are important additions to assessment protocols to establish the cognitive and behavioural implications of a child's executive difficulties in daily life (P. Anderson, 2002; Chan, Shum, Toulopoulou, & Chen, 2008). However, many tasks in the preschool-age range are aimed at measuring specific skills rather than EF in a more ecologically valid sense. In regards to intervention, it is important to understand what specific cognitive deficits are present, but to also gain an understanding of the amount and type of support a child requires in a typical activity(Henry & Bettenay, 2010). Tasks in everyday life require the integration of executive skills, unlike the artificial separation of cognitive domains that often occurs in neurocognitive assessments. Rocke and colleagues developed the Children's Kitchen Task Assessment (CKTA) to assess EF in a novel, but ecologically valid, task for eight to twelve-year-olds(Rocke, Hays, Edwards, & Berg, 2008). The task is novel in the sense that the child would not have completed this specific task before but is ecologically valid as it is similar in concept to tasks that a child would be expected to do in everyday life. This cue-based task requires children to follow step-by-step instructions with as little assistance as possible. It evaluates a child's EF and identifies how much support is needed for an individual child and when this support may be required. Using the CKTA, poorer EF was found in school-age children with sickle cell disease, a population with known EF deficits(Berg, Edwards, & King, 2012), with significant group differences observed for initiation, organization, and completion.

The aim of the two studies reported in this paper is to validate an ecological measure of executive functioning for use with preschool children and to use this measure to establish whether preschool children with SCA, a patient population with known executive difficulties in older children and adults, show differences at this earlier age.

Study 1: Validation and norming of the Preschool Executive Task Assessment (PETA)

The PETA was developed to measure EF in an ecologically valid way (Burgess et al., 2006; Schmuckler, 2001). Design requirements included a scoring system that is not dependent on linguistic ability or motor speed and accuracy, and a focus on process rather than accuracy so that a young child's executive skills can be appropriately captured. The PETA can be scored both quantitatively and qualitatively for different dimensions of interest and is not time limited. The purpose of a combined cueing system is to highlight a child's strengths and weaknesses as well as creating an objective way to reflect upon where a child's overall performance lies in comparison to their peers.

Method

Participants

Testing occurred in three nursery/school settings and at the London Babylab, UCL Institute of Child Health. Exclusionary criteria included a history of a developmental disorder and not being fluent in English, as determined by parental report. One hundred and sixty six children were recruited and assessed (mean age=4.5; range=3.0-6.0; 87 males). All children completed the PETA and WPPSI-III-UK. The supplementary section shows participant demographics and normative PETA task data (Table S3-S8).

Measures

Preschool Executive Task Assessment

The PETA was adapted from the CKTA, using a similar scoring and cueing system (Rocke et al., 2008). The task was designed so that a wide range of functioning can be accommodated, allowing for a more inclusive approach than a time limited or pass/fail task. The task involves using an "ingredients' box with pre-prepared materials, a recipe book, a timer, and cueing/scoring sheets (see Figure 1). The child follows a picture recipe book step-by-step, using the supplied materials, to make the final picture. The examiner delivers a pre- and post-task questionnaire, times task completion, and follows a cueing protocol (see Table S1). Children receive a total summary score (TS; a combination of amount and level of cues), as well as completion time, highest of five levels of support required during task, total number of cues required (TC), and cues required to initiate, sequence, and complete task. Qualitative scores for working memory, distractibility, organization, emotional control and self-talk are recorded by the examiner (see Table S2).

Figure 1. Illustration of systematically laid out "ingredient box" and child following recipe.

BRIEF-Preschool (Gioia, Espy, & Isquith, 2003)

The BRIEF-P consists of 63 items. It is comprised of five subscales that create three broader indexes Flexibility (FI), Emergent Metacognition (EMI), and Inhibitory Self-Control (ISCI) and a General Executive Composite (GEC). Higher scores indicate poorer EF (mean =50, SD=10).

Children's Behaviour Questionnaire-Short Version (Putnam & Rothbart, 2006)

The CBQ measures temperament in 3- to 7-year-olds using 13 subdomains. Two domains, attention and inhibitory control, are considered to be related to executive functioning.

Wechsler Preschool and Primary Scale: WPPSI-III-UK (Wechsler, 2002)

The WPPSI-III-UK is a standardized IQ measure used to obtain performance IQ (PIQ) and verbal IQ (VIQ). For three year olds, the full WPPSI form was administered but for four and five year olds, two verbal subtests (information and vocabulary) and two performance subtests (block design and matrix reasoning) were used to prorate PIQ and VIQ (Mean score =100, SD=15).

NIH Toolbox task of Inhibitory Control and Attention (Gershon et al., 2010)

In this standardised computerised measure, children are required to press the arrow button that corresponds to the same direction as the on-screen stimulus. Higher scores indicate better attention control (mean standardised score =100, SD=15).

Procedure

Ethical and R&D approval was obtained from the National Health Service and UCL Institute of Child Health. Written parental consent and child assent were obtained. Children were administered the WPPSI followed by the PETA. The subset of children in the laboratory setting also completed the NIH toolbox task of inhibition/attention. Parents/guardians filled out the CBQ and the BRIEF-P was completed by the classroom teacher or by the parent/guardian for the children who were tested at the London Babylab.

Statistical Analysis

Data analyses were conducted using SPSS for Mac version 21. The interclass correlation coefficient (ICC) was obtained for reliability analyses. A multivariate analysis of variance (MANOVA), chi-square and linear regression were used to look at the association between age and performance. Pearson's correlations were used to investigate associations between PETA performance and neuropsychological tasks. One-way ANOVA was used to investigate group differences based on qualitative ratings, and gender (male/female). Post hoc comparisons were conducted using Tukey HSD.

Results

Reliability

To test inter-rater reliability, ten testing sessions were coded by three testers with a strong inter-rater reliability (ICC =.93) and re-coded at least a week later showing evidence for strong intra-rater reliability (ICC= .88 to .98).

PETA Performance and Age

Age was strongly related to performance on all quantitative domains of the PETA, except for judgment/safety (Table 1). Differences in the TS score by age group (three, four, and five years) were investigated with univariate ANOVA. Post-hoc tests showed that performance significantly increased with age in line with the rapid development of executive skills reported during this period ($F_{2,164}$= 58.39, p <.001; figure 2). A linear regression found that chronological age predicted 40% of the TS ($F_{1,162}$ = 42.9, p <.001, R^2= .398). Younger children required higher levels of examiner support to complete the task with a chi-square showing that three-year-olds tended to require higher levels of cues (e.g. level 4-physical assistance; where the examiner completes part of a step when child does not respond to cues in level 3-direct verbal support) in comparison to four-year-olds with only a small number of five-year-olds requiring physical assistance (X^2 (8)=56.16, p <.001). Chi squares showed significant developmental trends for better performance with age in the qualitative domains of working memory (X^2 (4)=16.30, p =.003), organisation (X^2 (4)=12.97, p =.01), and emotional lability (X^2 (4)=11.77, p =.02), with older children receiving higher scores from the examiner for their performance in each of these domains. A non-significant trend for improvements in distractibility was observed (X^2 (4)=6.19, p =.15). (were the age results holding when you control for IQ?)

(TABLE 1 HERE)

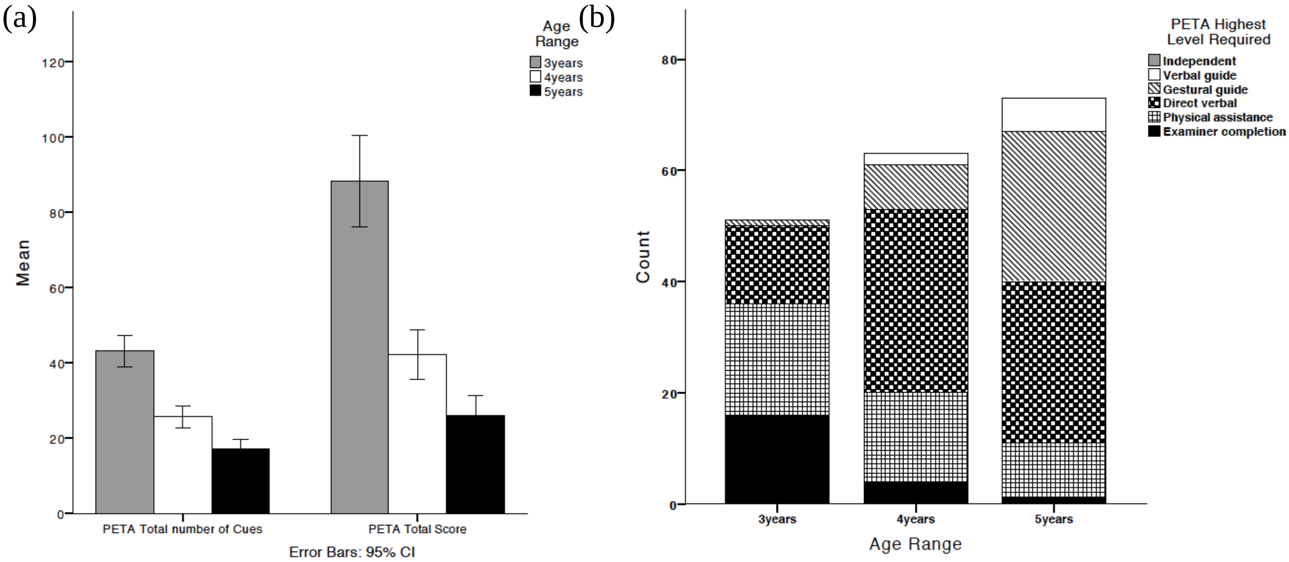

Fig 2. Typical Performance for (a) number of cues, total score (error bars = 95% confidence interval), and (b) highest level of support during task based on age range. Higher scores for number of total cues and the total score indicate poorer performance in 2(a).

PETA Convergent and Discriminant Validity

The convergent validity of the PETA was investigated by comparing quantitative scores with proxy-reported EF and the NIH toolbox task of inhibition/attention. Table 2 shows significant mild to moderate correlations between quantitative PETA scores and the BRIEF-P and CBQ executive domains (r=.18 to r=.31), providing evidence for the convergent validity of the PETA. Additionally, significant moderate to strong correlations were observed between PETA scores and performance on the NIH attention/inhibition (r=-.46 to r=-.60). Discriminant validity was investigated by comparing the PETA domains with the hypothetically unrelated constructs of the Block Design and Information WPPSI-III-UK subtests. Table 2 shows that some influence of WPPSI information and Block Design can be observed for PETA TC but the association with NIH attention/inhibition is stronger and significantly different from the correlation with Block Design (Z=-5.377, p=<.001) and Information (Z=-5.507, p=<.001). than the relationships observed for the executive tasks and is not observed for the other PETA domains evidencing the discriminant validity of the PETA. To further corroborate the discriminant validity of the PETA, the 'non-executive' domains of the CBQ were investigated and it was found that the associations observed for the inhibition and attention domains were not seen for the other domains.

(TABLE 2 HERE)

The qualitative PETA domains were investigated using one-way ANOVAs to determine whether the experimenter categorization of 'poor', 'typical', and 'very good' corresponded to teacher ratings on the approximate domains of the BRIEF-P. Although the model was not significant ($F_{3,96}$= 2.02, p =.14), examiner ratings of organization during the PETA task showed that, in general, the poor group obtained the poorest scores on the BRIEF-P Plan/organize domain, followed by the typical group, and the very good group. Distractibility categorization was compared with scores on the BRIEF-P Inhibit subdomain with a trend for mean scores going in the correct direction, but again the model was not significant ($F_{3,96}$=.348, p =.7). The model for working memory was also not significant ($F_{2,94}$= 1.26, p =.28), although the poor group had lower mean scores than the other two groups. There were no differences between the emotional lability groups ($F_{2,94}$= .045, p =.95) on the Emotional Control BRIEF-P domain (potentially due to the small number of children rated as poor; see Table 3 and the supplementary section).

Discussion

This study presents evidence for both the reliability and validity of the PETA as a useful tool for looking at executive development in both clinical and research settings. Rater reliability is high and task performance shows a significant improvement between three and five years in line with the current understanding of executive development in preschool-age children (Anderson, 2002). A limitation is the low to moderate correlations between the PETA domains and the proxy-report domains on the BRIEF-P and the CBQ; however this is typical for validation of similar measures in the literature (Dias & Seabra, 2012; Ponitz et al., 2008). Low correlations have previously been found between parent-report and performance based tasks(Mahone et al., 2002). To some degree, these correlations may be impacted by differences in task constructs and rater reliability for the BRIEF-P and the CBQ. However, the stronger association with the NIH task in comparison to the non-executive tasks corroborates the construct validity of the PETA. The quantitative measures were found to be more reliable than the qualitative measures. This may be due to the qualitative task domains measuring different aspects of EF constructs in comparison to the parent-report measure. However, the trend for children falling into appropriate categorical order is promising and the qualitative domains add to the informative quality of the assessment as they may capture EF aspects that are not commonly observed by parents. The PETA TS showed a greater developmental trend and stronger and more consistent relations with existing EF measures than it’s specific components. However, the real strength of this measure is the inclusion of these qualitative and quantitative components that allow for microanalysis to establish an individual child’s strengths and weaknesses. Further validation for the PETA (results in supplementary section) can also be observed in the relationship between self-talk and performance in the younger children(Winsler, Carlton, & Barry, 2000), the trend for girls performing slightly better than boys(Ponitz et al., 2008; Wiebe, Espy, & Charak, 2008) (Ponitz et al., 2008; Wiebe, Espy, & Charak, 2008), and the impact of poorer SES (Noble, Norman, & Farah, 2004).

(TABLE 3 HERE)

Study 2: Executive functioning in preschool children with sickle cell anaemia

A performance-based assessment that can inform a targeted intervention is an invaluable resource in the assessment of patients with potential EF deficits (Burgess et al., 2006). Berg et al. (2012) observed several differences between school-age children with SCD and a matched comparison group in task performance on an ecologically-valid task of executive functioning which led them to emphasise the importance of performance-based tasks to obtain a holistic picture of a child's ability. This study investigates whether executive deficits can be observed on an ecological level in preschool children with SCA. Earlier detection of potential executive deficits could lead to earlier interventions with the goal of improving school readiness and reducing the reported achievement gap in older patients(Schatz, 2004).

Method

Participants

Twenty-two patients with SCA between three and five years old whose parents identified themselves as Black British and had no history of neurological morbidity (Mean age 4.8, SD=.94; Mean IQ=98.6, SD=11.4; 13 males) were recruited at Barts NHS Trust and compared with 24 ethnicity, age, SES, and IQ-matched comparison children (Mean age 4.8, SD=.88; Mean IQ=101.5, SD=11.8; 10 males) that were recruited through the same clinics as the patients as well as schools in the same boroughs of East London where the patients reside.

Measures & Procedure

The PETA and the WPPSI-III-UK, as described in the methods section of Study 1, were administered to all patients and matched controls. The PETA always followed a break after the administration of the core subtests of the WPPSI-III-UK.

Statistical Analysis

Raw scores were converted to z scores based on the normative data obtained in Study 1. MANOVA, independent t-tests and chi-square analyses were used to look at group differences.

Results

A MANOVA on the quantitative PETA scores (Initiation, Sequencing, Meta-Cognition, Judgment/Safety, Completion) found a significant difference between the patients and the matched controls ($F_{1,45}$=2.5,p=.05). Inspection of individual quantitative subdomains revealed that the patients performed poorer on the domains of Completion and Sequencing (Table 4). A trend for poorer performance was observed for TC (t(44)=-1.6, p=.11), but not for Completion Time (t(44)=-.50, p=.62). Although non-significant, patients also had a higher mean TS, required more cues for the Meta-Cognition and Judgment/Safety domains. Chi-square analyses on the qualitative examiner-rated domains (Working Memory, Organisation, Emotional Lability, Distractibility) and Highest Level domain revealed significant group differences for Distractibility only (X=10.18, p=.002). A MANOVA on the quantitative PETA scores (Initiation, Sequencing, Meta-Cognition, Judgment/Safety, Completion) revealed that the patients were significantly different from the mean norms in Study 1 ($F_{5,178}$=10.42; p<.001), with significantly different means observed for Sequencing, Judgment/Safety and Completion. Chi-square analyses showed group differences for Distractibility only in the qualitative domains (X=5.7, p=.02). Independent T Tests showed a trend for a poorer TS in patients (t(184)=-1.8, p=.07) and significantly poorer performance for TC (t(184)=-22.2, p=.03) and Completion Time (t(184)=-2.6, p=.01) in the patient group.

(TABLE 4 HERE)

Discussion

It was expected that executive deficits would be observable on an ecological level in children with SCA. The patient group showed a trend for poorer performance in the TS and TC scores but there was no difference for Completion Time. There was a non-significant trend for poorer performance across the quantitative subdomains driven by poorer patient scores in the Completion, Sequencing, and Judgment/Safety domains. In the examiner-reported qualitative subdomains, there were differences observed for Distractibility only.

The lack of significant differences in TS and TC scores between the patients and the matched controls indicate that differences in EF in specific domains may not yet translate to differences in every day EF. The medium effect size for TC indicates that there was a tendency for patients to obtain more cues throughout the task. This meant that patients required more cues to stay on task and to provide scaffolding through the sequence of steps. Despite the lack of significant group differences on the macro level of the task, there were notable differences in specific quantitative and qualitative domains that were also observed when performance was compared with the normative scores.

Significant group differences were observed for Organisation, Initiation, and Completion in Berg and colleagues' (2012) cohort of eight to 12 year olds on the CKTA that uses similar scoring and cueing guidelines but only significant group differences for the Completion domain were found in the current cohort. Differences in study findings could be due to the differences in task design or could be a result of a younger and more homogenous population in the current study. Berg et al. included children who had a history of neurological morbidity and also included a patient with a different genotype whereas the current study excluded children with other sickle genotypes and children with a known history of neurological morbidity. However the shared group difference between studies for difficulties in task completion is particularly interesting as it is an aspect of EF research that has not previously been investigated in the sickle cell literature. Similar to the older children in the study by Berg et al., the patients in the current study also required more cues to complete the task. Maintaining intentions, or goal-directed behaviour, is the management of behaviour, including the activation and inhibition of actions, in order to reach goal completion, and is often impaired in disorders that affect the frontal lobes(Levine et al., 2000).

General Discussion & Future directions

There is a lack of validated EF measures for preschool children(Delis, Kramer, Kaplan, & Holdnack, 2004; Korkman, Kirk, & Kemp, 2007). Proxy-report screeners and specific executive tests that are widely used are limited in terms of gauging the impact of deficits on everyday life(Isquith, Crawford, Espy, & Gioia, 2005; Isquith, Gioia, & Espy, 2004). The authors are not aware of another performance-based ecologically valid task available for preschool children, despite the importance of EF for social skills and school readiness in this transition period(P. J. Anderson & Reidy, 2012). This task has the potential advantage over existing measures to provide a more holistic and multi-faceted picture of a child's EF(Espy, 1997).

Further research is required to establish whether the EF deficits observed for preschool children with SCA in the current study are replicated using other EF measures. A focus on early EF assessment could improve outcomes for young children with SCA by informing early interventions and reducing the reported achievement gap, particularly as executive problems are the most commonly reported deficits in older children with SCA (Schatz, Finke, Kellett, & Kramer, 2002). It could also indicate children who are most at-risk for future stroke as well as providing information for a baseline of ability for those children who later experience stroke(White et al., 2006).

Further work with other patient groups with known executive deficits is required to establish the utility of the PETA across different patient populations. Future research aims include the development of PETA profiles for clinical and research use as well as to develop comparable novel tasks for preschoolers that could be used as alternative versions of the PETA to maintain task novelty.

Key Points:

- There is a lack of executive assessments for preschool age children and no existing ecologically valid measures of executive functioning.
- We developed an ecological executive task with 166 three- to five-year-olds that adopts a micro-analytic approach to performance interpretation in order to identify where individual strengths and weaknesses lie in everyday tasks.
- Preschool children with sickle cell anaemia showed specific EF deficits on the ecological executive task.
- This measure is an important asset for clinicians who want to establish where a preschool child is struggling in every day tasks and to develop targeted interventions or support for children with executive deficits.

Acknowledgments:

The authors would like to thank the families and teachers who participated in this research. This research was funded by the Child Health Research Charitable Incorporated Organisation and supported by the National Institute for Health Research Biomedical Research Centre at

Great Ormond Street Hospital for Children NHS Foundation Trust and University College London. MdH was supported by Great Ormond Street Hospital Children's Charity. Task materials are available by request to first author.

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Table 1. Correlations between PETA domains and age

	PETA Domains							
	TS	TC	Initiation	Sequencing	Meta-cog	Judgment	Completion	Time
Age	-.631***	-.652***	-.431***	-.336***	-.559***	-.049	-.212***	-.481***

****p* <.005

Table 2. Correlations between quantitative PETA scores and behavioural reports on BRIEF-P and CBQ behavioural control domains

	PETA TS	PETA TC	Initiation	Sequencing	Meta-cog	Judgment	Completion	Time
BRIEF-P GEC	.181*	.196*	.280***	-.144	.141	.085	-.005	.183*
BRIEF-P EMI	.186*	.193*	.307***	-.099	.115	.101	-.028	.181*
BRIEF-P ISCI	.205*	.208*	.174	-.177	.186*	.128	.077	.192*
BRIEF-P FI	.152	.167	.174	-.183*	.109	.044	-.015	.181*
CBQ Attention	-.220***	-.224***	-.129	.205**	-.193*	-.122	-.023	-.172*
CBQ Inhibitory Control	-.187*	-.192*	-.086	.130	-.218***	-.091	-.052	-.229***

NIH attention and inhibition	-.554***	-.599***	-.474*	.265	-.337	-.458*	-.281	-.267
WPPSI Block Design	-.149	-.194*	-.174	.114	-.15	-.059	.000	-.132
WPPSI Information	-.159	-.186*	-.215**	.047	-.138	-.052	-.079	-.092

*p<.05 ** p <.01 *** p <.005

Table 3. Mean BRIEF-P subdomain scores divided into groups based on examiner rating during PETA administration as poor, typical, or very good on four categorical PETA domains (organisation, distractibility, working memory, emotional lability). There were no significant group differences on the BRIEF-P subdomains relevant to each PETA domain. However, there is a trend for mean scores going in the correct direction for the four domains (i.e. the poor groups obtaining the highest scores on the relevant BRIEF-P subdomains).

PETA Examiner Category Rating	BRIEF-P Plan/organize [a]	BRIEF-P Inhibit [b]	BRIEF-P Working Memory [c]	BRIEF-P Emotional Control [d]
Poor	53.2	48.6	54.9	46.8
Typical	48.3	47.9	49.4	46.6
Very Good	47.1	46.5	49.1	46.2

a As rated for the PETA Organisation domain b As rated for the PETA Distractibility domain

c As rated for the PETA Working Memory domain d As rated for the PETA Emotional Lability domain

Table 4. Group comparisons on the domains of the PETA

Variable **M (SD) /[range]**	**Patient (n=22)**	**Matched Controls (n=24)**	**P* (d)**	**London Mean Norms**	**P (d)**
Total Score	63.18 (48.2)	47.54 (31.2)	.195 (0.4)	46.6 (38.3)	.067 (.04)
Total Cues	34.32 (18.2)	26.71 (13.3)	.111 (0.5)	26.3 15.8	.028 (0.5)
Completion Time	16.2 (3.3)	15.5 (4.7)	.618 (0.2)	13.96 3.9	.011 (0.6)
Initiation	2.32 (2.7) [0-8]	2.33(2.4) [0-7]	.984	2.6 0-9	.623
Sequencing	.82 [0-3]	1.5 [0-5]	.034	1.87 0-7	.004
Meta-cognition	5.05 [1-9]	3.96 [1-9]	.132	4.27 0-9	.128
Judgment/Safety	1.18 [0-6]	.54 [0-5]	.122	.43 0-5	.044
Completion	2.77 [0-7]	1.21 [0-6]	.003	1.1 0-6	.001

Variable	Patient	Matched	P	London Mean Norms	P
%					
Working Memory			.123	*11*	*.196*
	13.6	4.5			
Poor	*68.2*	50		*51*	
	18.2	45.5			
Typical				*38*	
Very Good					
Organization		27.3	.924	*17*	*.538*
	22.7	31.8			
Poor	*36.4*	40.9			
	40.9			*38*	
Typical					
				45	
Very Good					
Emotional Lability		18.2	.645	*6*	*.223*
	13.6	54.5			
Poor	*68.2*	27.3		*62*	
	18.2				
Typical				*32*	
Very Good					
Distractability			.002	*13*	*.017*
Poor	*27.3*	0		*46*	
	54.5	45.5			

Typical	*18.2*	54.5		*41*	
Very Good					
Highest Level of Support			.825		*.551*
Verbal Guidance	*0*				
		0			
Gestural Guidance	*13.6*			*4.9*	
		20.8			
Direct Verbal	*36.4*			*19.5*	
		41.7			
Physical Assistance	*36.4*			*41.5*	
		29.2			
Examiner Completes	*13.6*			*23.2*	
		8.3			
				11	
Self-talk		47.4%	.485	*51.1*	*.561*
Yes	41.2%			*48.8*	