Temperament in preschool children with sickle cell anaemia

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What is known about this topic

- SCA can affect a number of cognitive and psychosocial outcomes
- There is an increased autonomic nervous system reactivity to stress in children with SCA which may have an influence on temperament

What this study adds

- Children with SCA experience more negative affectivity and higher rates of discomfort
- Discomfort is likely to be directly influenced by the physical sickle cell disease process, reflected here by the number of hospital admissions.
- Lower internal consistency of the CBQ for children with SCA should be considered in future research.

ABSTRACT

Aims: Few studies have investigated the potential impact of sickle cell anaemia (SCA) on temperament. The aim of the current study was to investigate temperament in preschool children with SCA and to establish the reliability of the Children's Behaviour Questionnaire (CBQ) in this population. Methods: The CBQ, a parent-report measure of temperament was completed by parents of 21 preschool children with SCA and a control group of parents of typically developing children, matched for age, ethnicity, and socio-economic status. Results: A significant difference between groups was identified in relation to the dimension of Negative Affectivity only, with specific differences observed in the Discomfort subdomain. Low alpha values were observed on multiple subdomains. Conclusions: Patients are reported to have higher rates of Negative Affectivity, particularly Discomfort. Lower internal consistency of the CBQ for children with SCA should be considered in future research.

INTRODUCTION

Sickle cell disease (SCD) is a group of genetic disorders, characterised by the presence of HbS and another abnormal variant of haemoglobin. The primary physical

symptoms of SCD begin to emerge at around six months old, with severity of symptomatology varying, but including acute pain, increased risk of infection and pulmonary, respiratory and cardiac complications, as well as potential cognitive issues (1). Sickle cell anaemia (SCA) is the most severe and common form of SCD, causing blood cells to change from their regular shape to a crescent or "sickle" shape. Temperament is one of the many developmental outcomes that may be affected by a child's experience of chronic illness. Temperament can be described as individual differences in emotional, motor and attentional response to events and the way in which an individual modulates or self-regulates their reactions (2). It is typically characterised as a predisposition to respond to the environment in certain ways and it is hypothesised to emerge at an early developmental stage. It has been postulated that increased autonomic nervous system reactivity to stress in young children with SCA may have an influence on temperament development (3). Only one study has investigated the potential impact of SCD on temperament development in young children, however they did not include a typically developing control group. They reported that toddlers with SCA displayed less activity than children with less severe SCD genotypes (3). The only other study to investigate temperament in SCD reported no differences in adaptability, attention focus, or general rhythmicity in 32 school age children with SCD (4). Both studies combined the findings for children with SCA with other sickle genotypes so potential differences for children with SCA, typically the most severe form of SCD, remains unknown.

The present study investigates potential parent-reported differences in temperament on the Children's Behaviour Questionnaire (CBQ) in preschool children with SCA, in comparison to typically developing controls matched for ethnicity, age, and SES. The

CBQ, a reliable assessment of reactive or self-regulative behaviours in preschool children (5), is frequently used to measure temperament in 3 to 8 year olds (2). A strong relation between the development of self-regulation and maladaptive temperament on the CBQ and problem behaviour in preschool children highlights the importance of assessing temperament in paediatric patient populations (6). SCD is associated with increased anxiety, depression, behavioural and social issues, as well as poorer quality of life, and so a better understanding of early adaptive responses in children with SCA is required (7). Research indicates that children with chronic pain tend to demonstrate more difficult temperaments than their peers (8,9). Furthermore, children with more difficult temperaments tend to be more reactive to stress and more sensitive to pain than other children (10).

Temperament has been shown to predict psychological adaptation to diagnosis and treatment in childhood disorders. For example, temperament on the CBQ can predict paediatric cancer patients' quality of life (11). The relation between specific temperament dimensions and higher reports of pain has also been documented with paediatric cancer patients (12). Therefore, assessing temperament early in treatment may help better identify children who are at a higher risk for poor quality of life. In addition to investigating the temperament of preschool children with the CBQ, the current study will also investigate the internal consistency of this commonly used developmental scale to establish the reliability of this measure in young children with SCA.

METHOD

Procedure

Children with and without SCA were recruited for this case-control study. Ethical approval was obtained from the local NHS ethics committee and University College London Great Ormond Street Institute of Child Health. Parents of patients who met the study criteria were informed of the project during routine clinical visits at Royal London Hospital. Written informed consent was obtained. The short version of the CBQ was administered to parents to assess temperament (2). A one-way MANOVA was performed to examine whether overall temperament differed between groups. Cronbach's alpha was used as the reliability analysis to investigate the internal consistency of the CBQ subdomains in children with SCA.

Measure

The CBQ questionnaire consists of 94 statements that form three temperament dimensions of Surgency, Effortful Control and Negative Affectivity comprised of 15 subdomains including impulsivity, inhibitory control, focusing attention, shyness, sadness, anger/frustration, and activity levels. Parents are asked to answer, on a scale of 1 (extremely untrue) to 7 (extremely true), how true each statement is for their child.

Participants

Parents of patients with SCA aged 3 to 5 years with no history of neurological issues were recruited. Control children, matched for ethnicity, age, and socio-economic status (SES, by postcode), were recruited from local boroughs.

RESULTS

Parents of 22 Black British patients with SCA (*mean age* = 4.76) were recruited. Of these, 21 parents successfully completed the CBQ. All parents of 26 control children (*mean age* = 4.91) successfully completed the CBQ.

Scale Reliability in SCA

Alpha scores were generally lower in relation to the group of children with SCA than the values reported for typically developing children, with many not meeting the generally accepted value of .65 (1) (Table 1). Notably, the scores were also lower in the current control group, which may be a result of factors, such as SES, that this group were matched for.

Group Differences

There was no significant difference in overall temperament between children with SCA and controls; F (15,31) =1.21, p=.32, Wilk's ^ = .63. Given the variance in alpha values across subdomains, temperament on the three CBQ dimensions and individual subdomains were further investigated. Of the three CBQ dimensions – Surgency, Effortful Control and Negative Affectivity – the only significant difference between the groups was observed in relation to Negative Affectivity; F (1,45) = 12.22, p = .001 with the patient group displaying higher scores (M = 4.23, SD = .58) than the children without SCA (M = 3.65, SD =.56). Further analysis of the 15 subdomains of the CBQ showed that there was a significant difference between the two groups for discomfort only (Table 2). This discrepancy was further investigated to explore whether parent-reported discomfort was related to disease severity (defined as number of hospital admissions in the previous year for the current study). Higher discomfort

on the CBQ was associated with the number of hospital admissions in the previous year for the children with SCA (r=.567, p=.007).

DISCUSSION

This is the first study to investigate temperament in young children with SCA in comparison to matched controls. The participants with SCA scored significantly poorer on the discomfort subdomain and the negative affectivity dimension, characterised by dysregulated negative emotions, than the matched comparison group. Previous research in children with SCD has shown that more negative emotions were associated with increased pain and less participation in daily activities (13). Higher scores in discomfort and perceptual sensitivity for the patient group could be due to a higher reactivity to pain as a result of the sickle cell disease process (14). This is in line with previous research in toddlers with SCD that reported lower activity in young children with SCD, a finding that also could be attributed to the disease process (3).

The only previous study to explore differences between children with SCD and matched peers purposefully excluded items relating to the potential medical effects of SCA, such as overtiredness (4). The lack of significant differences in other temperament dimensions reflects the findings of the only other study to explore temperament in young children with SCA (3). The current study found that higher discomfort was related to number of hospital admissions for the patient group, however the potential influence of silent cerebral infarcts is unknown as these patients did not have MRI scans. Previous research has found no relation between a greater number of admissions (or painful events) in children with SCA and a higher rate of silent cerebral infarcts (15).

The CBQ is widely used in the preschool literature; however, to the author's knowledge it has not been previously administered to young families with SCA. Although poorer internal consistency has been found in African American and lower SES families, the alpha coefficients for internal consistency are generally stronger for the subdomain of discomfort in the short scale than the standard scale (2), the only domain that was significantly different between the two groups in the current study. Nevertheless, lower alpha scores for the CBQ domains in children with SCA in the current study should be considered in future research with this patient population. Notably, low alpha values were also observed for the matched comparison group, which is in line with reports of poorer scale reliability in lower SES and ethnic minority families (2). The small patient population limits the generalisability of current study findings; however, results are strengthened by the inclusion of a matched control group

In conclusion, the children with SCA in the present study show differences for discomfort and negative affectivity when temperament is compared with matched peers. Levels of discomfort and negative affectivity may be directly influenced by aspects of the physical sickle cell disease process, specifically the experience of pain, as supported by the observed association with number of hospital admissions. These temperament traits require further investigation and due consideration in the clinical management of children with SCA.

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Table 1. Comparison of Cronbach's Alpha for the CBQ-short in children with SCA

Domain	**Putnam & Rothbart (2006) N=1,189**	**Current SCA Group N=21**	**Current Matched Group N=26**
Activity Level	.75	.46	.39
Anger/Frustration	.76	.33	.71
Approach/Positive	*65*	*.12*	*.64*
Attentional Focusing	*.75*	*.40*	*.20*
Discomfort	.79	.57	.59
Soothability	.73	.58	.11
Fear	.68	.36	.41
High Intensity Pleasure	.72	.68	.38
Impulsivity	.72	.77	.79
Inhibitory Control	*.72*	*.11*	*.12*
Low Intensity Pleasure	*.69*	*.77*	*.61*
Perceptual Sensitivity	.73	.25	.65
Sadness	.61	.37	.45
Shyness	.85	.64	.84
Smiling/Laughter	.71	.35	.60

Table 2. Group differences on the individual subscales of the CBQ-S using one-way ANOVA.

	Patients (N=21)	Ethnicity Matched	Group Effect

	Mean (SD)	Controls (N=26) Mean (SD)	F	p
Activity Level	4.6 (0.93)	4.9 (0.75)	1.225	0.274
Anger/Frustration	4.4(1.02)	3.9 (1.2)	2.42	0.127
Approach/Positive Anticipation	5.3 (0.82)	5.0 (0.92)	1.01	0.321
Attentional Focusing	4.9 (1.04)	4.9 (0.89)	.003	0.953
Discomfort	5.14 (0.99)	4.25 (1.02)	10.294	0.002**
Falling Reactivity/Soothability	5.1 (0.99)	5.5 (0.65)	2.931	0.094
Fear	4.4 (0.96)	3.9 (1.11)	3.708	0.060
High Intensity Pleasure	4.42(1.06)	4.62 (0.84)	.411	0.525
Impulsivity	4.3 (0.67)	4.4 (1.01)	.183	0.671
Inhibitory Control	5.0 (0.99)	4.7 (0.61)	.785	0.380
Low Intensity Pleasure	6.1 (0.73)	5.9 (0.67)	.335	0.566
Perceptual Sensitivity	5.8 (0.66)	5.33 (0..92)	3.013	0.089
Sadness	4.4 (1.17)	3.9 (0.87)	2.243	0.141
Shyness	3.9 (1.2)	3.2 (1.3)	2.884	.096
Smiling/Laughter	5.6 (0.77)	5.8 (0.85)	.375	0.543

***p<.001 ** p<.01 *p<.05