



Cognitive Effects of Sickle Cell Disease in Children

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Introduction

- Sickle Cell Disease (SCD) is a blood disorder that leads to malformed blood cells and blood flow obstruction.
- SCD evidences diverse adverse cognitive sequelae (e.g., Armstrong et al., 1996; Berkelhammer et al., 2007; Connolly et al., 2019; Kawadler et al., 2016; Kwiatkowski et al., 2009; Wang et al., 2001)
- There is significant variability of severity level and disease presentation, making it a complex task to study cognitive impact (e.g., Lance et al., 2015; Prussien et al., 2020; Steen et al., 2003).
- This research aimed to examine the specific cognitive domains impacted by disease severity and age.

Hypotheses:

- Cognitive deficits in children with SCD will vary by disease severity, with greater impairment in visual-spatial domains
- Decline in cognitive functioning will increase with age.

Methods

- Data from the Cooperative Study of Sickle Cell Disease was analyzed. Patients ($n = 2408$) with SCD and 14 years of age and younger were included in the current study.
- Sample predominately identified as Black or African American (96.9%, $n = 2334$).
- Measures included Wechsler Intelligence Scale for Children (WISC-R/WISC-III), Woodcock Johnson-Revised (WJ-R), and information from medical records.
- A one-way ANOVA assessed the relationship between cognitive scores and disease subtypes. A Pearson R Correlation was used to evaluate the relationship between age and cognitive performance.

Results

Disease severity was negatively correlated with some aspects of cognitive functioning, specifically in the visual-spatial domain [$F(2, 1) = 4.42, p = .013, \eta^2 = .02$].

WISC Indices and Subtests	F	P-value	WJ Cluster/Subtests	F	P-value
Full Scale IQ	1.039	.355	Broad Reading	.970	.380
Verbal IQ	.532	.588	Letter Word	1.342	.263
Performance IQ	1.327	.266	Word Attack	.317	.729
Verbal Comprehension	1.361	.269	Passage Comprehension	.563	.570
Perceptual Organization	.977	.386	Broad Math	2.714	.068
Freedom from Distractibility	3.317	.048	Calculation	4.833	.008
Processing Speed	3.545	.040	Applied Problems	1.441	.238
Picture Completion	1.449	.236	Processing Speed	2.306	.114
Information	.996	.370	Visual Matching	1.853	.170
Coding	1.629	.197	Cross Out	1.475	.242
Similarities	.829	.437	Short Term Memory	.755	.477
Picture Arrangement	.088	.916	Memory for Sentences	1.240	.300
Arithmetic	1.985	.139	Memory for Words	.129	.879
Block Design	4.422	.013	Analysis/Synthesis	1.823	.175
Vocabulary	.913	.402	Spatial Relations	1.510	.235
Object Assembly	.528	.590	Visual Motor Integration	.567	.572
Comprehension	.424	.655			
Symbol Search	1.624	.212			
Digit Span	.815	.445			

Pearson R Correlation for Age and WISC

		Correlations											
		CURRAGEY	NPSCFSIQ	NPCIMQ	NPCIPIQ	NPCIVC	NPCIPQ	NPCIFD	NPCIPS	NPCSPIC	NPCINFO	NPCSCOD	NPCSSIM
CURRAGEY	Pearson Correlation	1	-.026	-.044	-.037	-.257	-.356	-.028	-.136	.062	-.111	-.115	.160
	Sig.		.599	.371	.447	.096	.019	.861	.404	.203	.023	.019	.001
NPSCFSIQ	Pearson Correlation		1	.905	.878	.546	.549	.733	.653	.661	.741	.490	.704
	Sig.			.000	.000	.000	.000	.000	.000	.000	.000	.000	.000
NPCIMQ	Pearson Correlation			1	.599	.602	.399	.753	.475	.471	.808	.299	.785
	Sig.				.000	.000	.007	.000	.001	.000	.000	.000	.000
NPCIPIQ	Pearson Correlation				1	.385	.617	.564	.738	.720	.505	.603	.455
	Sig.					.009	.000	.000	.000	.000	.000	.000	.000
NPCIVC	Pearson Correlation					1	.822	.650	.461	.228	.293	.383	.403
	Sig.						.000	.000	.132	.051	.009	.006	
NPCIPQ	Pearson Correlation						1	.547	.583	.437	.178	.482	.262
	Sig.							.000	.000	.003	.243	.001	.082
NPCIFD	Pearson Correlation							1	.442	.485	.700	.414	.562
	Sig.								.003	.001	.000	.005	.000
NPCIPS	Pearson Correlation								1	.459	.374	.870	.392
	Sig.									.002	.015	.000	.010
NPCSPIC	Pearson Correlation									1	.389	.232	.359
	Sig.										.000	.000	.000
NPCINFO	Pearson Correlation										1	.234	.547
	Sig.											.000	.000
NPCSCOD	Pearson Correlation											1	.199
	Sig.												.000
NPCSSIM	Pearson Correlation												1
	Sig.												

Age was inversely correlated with measures of perceptual organization ($r = -.36, p = .02$) and visual motor abilities ($r = -.49, p = .001$).

Conclusions

- Results revealed that disease severity was negatively correlated with visual-spatial tasks.
- Decline in cognitive performance increased with age on tasks of perceptual reasoning and visual motor integration skills.
- Introducing remediation interventions targeted at these particular domains at earlier stages may help to reduce significant cognitive deficits.

