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EDUCATION

Reading intervention targeting phonemic awareness and symbol imagery in children with sickle cell disease

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Abstract

Children with sickle cell disease (SCD) frequently have diminished academic attainment and are particularly vulnerable to reading dysfunction. We explored the effectiveness of a multisensory reading intervention offered during the summer to children with SCD at our institution. Subjects with reading deficits were identified through parent report, clinical findings, or school meetings. Summer reading programs utilizing Phonemic Awareness and Symbol Imagery were provided. The Lindamood–Bell Auditory Conceptualization/Phonemic Awareness Test, Third Edition (LAC-3), and the Symbol Imagery Test were used as pre- and postintervention examinations to measure progress. Fifteen students (median age 9.4 years, range 6–14 years, eight females, all African American) received the Phonemic Awareness intervention, two times a week for 6 weeks. The subjects showed statistically significant gains in standard scores derived from the LAC-3 (mean change 7.9 points, $p < .001$), with associated improvements in age equivalency (AE) and grade equivalency (GE). Twenty-nine students (median age 9 years, range 6–17 years, 13 females, all African American) participated in the Symbol Imagery reading program, also two times a week for 6 weeks. These students showed significant gains in overall standard scores (mean change 9.8 points, $p < .001$). Although results should be interpreted with caution due to small sample sizes, we found that summer reading clinics for children with SCD improved phonological processing and symbol imagery skills, potentially leading to substantial gains in reading capability.

KEYWORDS
phonemic awareness, reading, sickle cell, symbol imagery

1 | INTRODUCTION

Sickle cell disease (SCD) affects approximately 30,000 students in the United States. Although the medical treatment options available for individuals with SCD have improved dramatically, the direct and indirect effects of this condition, which negatively influence neurocognition and academic performance, have not been fully examined and few educational interventions have been created.

The hallmark of SCD is the painful vaso-occlusive event, but central nervous system effects are extremely frequent and include clinically apparent overt stroke and silent cerebral infarction detected through neuroimaging. Brain pathophysiology intertwined with social and environmental risk factors commonly cause neurocognitive delay.[1,2] Neurodevelopmental difficulties, identified through comprehensive

Abbreviations: AE, age equivalency; GE, grade equivalency; LAC-3, Lindamood–Bell Auditory Conceptualization/Phonemic Awareness Test, Third Edition; LiPS, Lindamood Phoneme Sequencing Program for Reading, Spelling, and Speech; SCD, sickle cell disease; SD, standard deviation; STARR, Success Through Academic Resources and Research.

neuropsychologic evaluations, begin in infancy or early childhood and may result in long-lasting detriment to academic achievement.[1–6] The chronic anemia of SCD resulting in diminished tissue oxygenation has been implicated in causing neurocognitive delay. In addition, children with SCD tend to live in socioeconomically disadvantaged neighborhoods and households, resulting in environmental factors that are highly associated with risk for poorer cognitive and academic performance.[7,8] Approximately 18% of students with SCD are retained in grade at least once,[9] and 30% of individuals with SCD fail to graduate from high school.[7] The medical and socioeconomic barriers that children and adolescents with SCD face increase the risk that they will lack the appropriate academic and functional skills needed to transition successfully to adulthood.[7,10]

The National Institute of Child Health and Human Development (NICHD) determined through a longitudinal study that about 40% of the general population had problems with reading that were severe enough to hinder their enjoyment of that activity.[11] Intervention through early direct instruction is considered best for these problems, as reading is a learned behavior rather than a developmental issue.[12] Children who fall behind in the kindergarten or first grade years generally fall further behind over time. Although children from low-income backgrounds may make gains in reading skills consistent with their peers during the school year, they demonstrate greater setbacks in reading skills during the summer.[13] However, summer reading programs have consistently demonstrated greater improvement in reading skills in low-income students compared to middle- or high-income peers.[13,14]

The most reliable indicator of a reading disability is an inability to decode single words, which can be determined by measuring a child's listening comprehension skills and which is negatively affected by deficits in the auditory discrimination domain.[11] In a study evaluating readiness for kindergarten in children with SCD, the Developing Skills Checklist was compared to those of a matched control group of kindergarten students in the Memphis city schools.[15] Scores were significantly lower in children with SCD than controls in the domain of auditory discrimination. This suggests a substantial barrier for the academic success of SCD students, because auditory discrimination is needed for acquiring phonemic awareness, enhancing symbol imagery, and, eventually, reading.[16] In fact, lack of phonemic awareness and inability to use symbol imagery have been shown to be major obstacles for learning to read.[17]

At St. Jude Children's Research Hospital, the STARR (Success Through Academic Resources and Research) program provides educational support for children with SCD and is staffed by educators/teachers working with the hematology department. The STARR program assists patients in three primary ways. Initially, the educators complete a consultation with families in clinic to identify school problems and academic difficulties. If problems are identified, the teachers meet with other professionals and provide referrals within the hospital and community (e.g., psychology, social work, speech therapy, Tennessee Early Intervention Services). In addition to helping families acquire resources in the community, the educators of the STARR program visit the patient's community school to provide education about SCD and advocate for the academic services available for students with disabilities. Lastly, teachers in the STARR program design and facilitate programs to meet patient needs, such as the Hematology Teen Support Group.

The STARR program also established a summer reading program for children with SCD, which focused initially on improving reading skills through enhancing phonological processing and subsequently through the addition of symbol imagery. The goals of this intervention were (a) to provide a supplemental summer reading program for children with SCD who had experienced reading difficulties, and (b) to obtain preliminary information regarding the short-term efficacy of these interventions.

2 | METHODS

This study utilized data collected in pre- and postintervention assessments administered to students with SCD who completed a reading intervention summer program. Participants in this program were identified through various sources. A psychological test, which demonstrated reading deficits, was often a source of referral. The most common one was the Woodcock Johnson III Letter-Word Identification Test. In some cases, a teacher reported a child having problems with reading during a meeting with STARR program educators to discuss an individualized education plan or a Section 504 Plan of the Rehabilitation Act of 1973. In addition, problems with their child's reading sometimes were reported by parents during a regular sickle cell clinic visit. In general, families of patients with known reading problems were informed of the existence of the program and encouraged to participate.

The Summer Reading Clinic utilized an interdisciplinary team to create an individualized curriculum for each student. Although the clinic was planned and facilitated by academic coordinators (educators) from the STARR program, input was included from social work and the hematology and psychology departments. This helped the team address the requirements of a patient population with diverse academic needs and psychosocial problems. The academic coordinators were professional educators licensed in the state of Tennessee. They had developed a strong trusting relationship with the students and caregivers before the beginning of the summer reading program through seeing them frequently during hematology clinic visits and helping them by advocating in the patient's home school and community. In general, the academic coordinators in our program had personal knowledge of each family's psychosocial and medical histories and helped coordinate care with the hospital psychosocial staff.

When a child was identified for the summer reading clinic, a packet that included a cover letter verifying days and times, a parent agreement, an absenteeism policy, and information about the content of the reading program was provided to the family. Students were assigned to programs based on needs identified in the assessment before reading clinic officially began. The Phonemic Awareness program was offered for students working on basic concepts like identification of letters and sounds. However, many patients did not require the program for

Phonemic Awareness, but needed to focus on concepts in the Symbol Imagery program.

On the first day of the reading clinic, the child's initial testing (either LAC-3 or Symbol Imagery test) was performed, while the parent waited in the reception area. The Phonemic Awareness program sessions lasted for 1.5 hours and were held twice a week for 6 weeks; the Symbol Imagery sessions were similar in length. Students were tutored in a one-to-one manner. Participants engaged in social breaks between their lessons, utilized mindfulness strategies and journaling, and were encouraged with small prizes from the class "treasure chest." Only limited observation of the sessions by a parent was allowed at the beginning, because students tended to be inhibited when their parent was watching. At least a 5-minute session of verbal communication was given to parents regarding progress for each session. Parents were also taught strategies and skills to work on at home to address the child's weaknesses. Skills were not standardized, but were tailored specifically to address the student's weaknesses. For example, for the children struggling with phonemic awareness due to trouble with mental imagery, students would learn to "air-write" or "air-draw" words by holding up their finger and pretending to create letters in the air. (This strategy is helpful for a patient studying for a spelling test, but also for other important visualizing skills, such as memorizing countries for a map test.) At the end of the program, parents were provided a packet of materials used during the clinic and another opportunity to meet with the teacher. Materials given to families were customized to the needs of the patient. Children would be given packets that included materials such as a whisper phone, felt squares for manipulating sounds, a pack of cards with sight vocabulary words to practice, books to read at home with parents, and journals for completing writing prompts and reflections on the program. The student's data also were shared with the child's school to assist in academic planning for the school year.

Phonemic awareness is the ability to auditorily perceive the identity, number, and sequence of sounds within words.[18] The Lindamood Phoneme Sequencing Program for Reading, Spelling, and Speech (LiPS) was utilized as the first reading-improvement method.[19] The Lindamood–Bell Auditory Conceptualization/Phonemic Awareness Test, Third Edition (LAC-3) was used to collect pre- and postintervention data. This standardized instrument is a norm-referenced and validated measure of auditory perception and conceptualization of speech sounds. The LAC-3 includes five categories: *Isolated Phoneme Patterns*, *Tracking Phonemes (Monosyllables)*, *Counting Syllables (Multisyllables)*, *Tracking Syllables (Multisyllables)*, *and Tracking Syllables and Phonemes (Multisyllables)*. The student is checked on conceptualization of five types of contrast: additions, shifts, substitutions, omissions, and repetitions. Raw scores on the LAC-3 are converted into age-based standard scores that have a mean of 100 and a standard deviation (SD) of 15. Age and Grade Equivalent (AE and GE) scores indicate the age and grade levels that correspond to a raw score. Because interpolation, extrapolation, and smoothing were used to create AE and GE, and their distributions were not normalized, these scores are meant to apply to single individuals only and aggregated statistical analysis is not reliable.

Symbol imagery is "the ability to create mental representations (i.e., imagery) for the sounds and letters within words" and is described as a sensory-cognitive function underlying phonological and orthographic processing.[20] The concept of symbol imagery was developed with recognition that phonemic awareness and improved "word attack skills," although important in the reading process, were often not enough to achieve successful literacy, particularly in the areas of word recognition and paragraph reading. Orthographic processing has been described as "representing a printed word in memory and accessing the whole word unit …or a letter cluster unit" and has been linked to reading comprehension.[21]

The Symbol Imagery Test assesses both phonological and orthographic skills needed for reading and spelling and indicates whether a student needs remediation of a symbol imagery deficit.[20] This standardized norm-referenced test is designed to measure a student's imagery for letters in both random and orthographically regular combinations from both visual and auditory stimulation. For each item, the examiner holds up a card for 3 seconds and then takes it away and asks the student what was seen. The test provides raw scores, standard scores, percentile ranks, and age equivalents. The evaluation includes five subsections: (a) student sees and recalls unconnected letters; (b) student sees and recalls nonwords; (c) student hears and spells nonwords; (d) student sees and manipulates one-syllable nonwords; and (e) student sees and manipulates multisyllable nonwords. Each section includes eight to 12 questions. "Manipulation" includes questions/requests such as "what is the fifth letter you saw" and "say the letters backwards."

2.1 | Statistical analysis

Standard scores of patients who participated in the Phonemic Awareness and Symbol Imagery programs were reported using means and SDs. Paired differences between post-test and pre-test standard scores were assessed with a paired *t*-test because the Shapiro–Wilk test showed normal distribution. For the Symbol Imagery program, Spearman correlations were used to test correlations between participant age and pre- and postintervention scores and pre-/postintervention score differences. *p*-Values were two-sided and $p < .05$ was considered significant. All analyses were conducted in SAS 9.4.

3 | RESULTS

3.1 | Phonemic Awareness

Fifteen patients completed the Phonemic Awareness program. Their median age was 9.4 years, with a range of 6.1–13.8 years. Ten patients (67%) had HbSS, three HbSC, and two HbS/β^+ thalassemia. Eight subjects (53%) were female; all were African American. Although neuropsychological testing was not required or standardized, the most relevant and commonly performed exam was the Woodcock Johnson III Letter-Word Identification Test. Among 10 participants that completed this test, the median standard score was 85, with a range of 71–104.

In the evaluation of phonemic awareness, the mean (±SD) pre-program and post-program standard scores on the LAC-3 were 93.4 (±8.5) and 101.3 (±10.0), respectively. Mean change in the standard score was 7.9 (±6.5) ($p < .001$). Improved individual AE and GE scores were observed, but these data could not be aggregated for the reasons indicated in Methods section. Tracking phonemes (monosyllables) and tracking syllables (multisyllables) were subsections in which most subjects demonstrated improvement.

3.2 | Symbol Imagery

Twenty-nine patients completed the Symbol Imagery program. The subjects had a median age of 9 years, with a range of 6–17 years; 13 (43%) were female and all were African American. Mean (±SD) pre-program and post-program standard scores were 86.6 (±20.1) and 96.4 (±14.4), respectively. Twenty-five of the 29 (86%) had increases in their standard score following participation. The average improvement in standard score was 9.8 (±13.6) ($p < .001$). Scores were increased more often than decreased in all five subsections, with the largest improvements in Subsection C (student hears and spells nonwords). Postintervention standard score performance was consistently better than preintervention performance, and differences in scores were not correlated with age of the subject.

4 | DISCUSSION

In children with SCD, deficits in cognitive development, particularly in language processing, have been reported in several studies[5,15] and in part reflect socio-environmental factors.[8] Unfortunately, interventions to alleviate this impairment in sickle cell patients have been almost nonexistent. Our academic coordinators' summer reading program has demonstrated for the first time that it is possible to reduce the reading deficits commonly found in children with SCD.

In 2000, The National Reading Panel released the findings of a longitudinal study, which recommended five areas of focus to improve reading in the United States: phonemic awareness, phonics, reading fluency, vocabulary development, and reading comprehension.[22] Explicit and systematic instruction in phonics teaches a fundamental reading skill that all students need to gain access to self-correction and to advance in the reading process (spelling and pronunciation). The "Nation's Report Card" noted that only 37% of 12th graders read at or above a proficient level.[11] Also concerning was that only 17% of African American students were in the proficient group, compared with 46% of White students, 25% of Hispanic students, and 49% of Asian students. Recent reports of intensive summer reading intervention for children with reading disabilities have indicated improvements in word and pseudoword reading, symbol imagery, and oral reading fluency in a cohort of 6–9-year olds[23] and decoding, oral reading skills, and comprehension in 6–12-year olds.[24]

Rather than targeting other areas of reading, such as vocabulary development, fluency, or comprehension, our program directed attention to phonological processing and subsequently to symbol imagery. We sought to address the reported need for interventions to alleviate the compromised academic performance described in the SCD literature.[15,25] In addition, we recognized that children with chronic illnesses and cognitive disabilities frequently have more adverse school experiences, and experience stigmatization associated with their disability. Struggling readers often face crippling loss of confidence in their abilities, are used to performing poorly in school, and may experience grade retention. Because the academic coordinators in our program had personal knowledge of each family's psychosocial and medical histories, and because they coordinated care with the hospital psychosocial staff, they were relatively well equipped to address psychosocial problems and/or minor acute medical issues during the reading program instruction, which led to a more pleasant environment for the students.

Our clinic used a reading methodology founded on auditory conceptualization by C.H. and P.C. Lindamood, who also coined the term "phoneme awareness."[19] They defined auditory conceptualization as "the ability to encode, or visually represent, precisely how and where two spoken patterns vary in the number, identity, and sequence of their sounds." Students who have difficulty in identifying phonemes within words will not understand the structural connections between the sequence of phonemes and the sequence of letters, and will have to rely on rote memorization to read and spell. Problems in executive function and attention along with delays in vocabulary, comprehension, and general verbal ability are observed in patients with SCD.[26,27] Deficits in working memory and auditory processing skills have also been described.[16] A broader examination of language processing delays in young children with SCD found specific problems in phonological skills.[25]

Beyond phonemic awareness, children must create mental imagery for the sounds and letters within words (symbol imagery). As summarized by Nanci Bell, "the ability to visualize letters within words is an important aspect of reading and spelling. Symbol imagery underlies both phonological and orthographic processing and, consequently, fluent, self-correcting reading behaviors."[20] In our summer reading program, we focused entirely on phonemic awareness initially, but in subsequent summers added the component of symbol imagery.

The improvements seen in our children with SCD after instruction in Phonemic Awareness and Symbol Imagery education were significant. These results were notable because of the speed with which the gains occurred. For example, participants across the school age spectrum from 7 to 17 years of age demonstrated a nearly two thirds of an SD improvement in standard scores after 6 weeks in response to the Symbol Imagery program.

However, our results should be interpreted cautiously because of several limitations. Our sample sizes were relatively small and the data were examined retrospectively. In part because the reading program was initiated as a "service" to our sickle cell patients, we did not have a control group of students who were evaluated without a reading intervention. Some students were unable to complete the reading program because of issues with illness, transportation, family vacation, and so forth. Only a limited number of students had sequential experience

with both reading interventions. We did not examine broader reading outcomes, such as word recognition, reading fluency, or reading comprehension. Unfortunately, although the teachers had hoped to follow the students throughout the school year to extend their work, the reach of the program was limited due to staffing needs. Throughout the school year, the educators gave primary attention to advocating for patients in their community schools. Therefore, we did not have follow-up information regarding school performance in reading in the years following a summer intervention.

In conclusion, we established initial efficacy of a summer reading intervention for children with SCD. Both Phonemic Awareness and Symbol Imagery reading interventions resulted in significant gains. The summer reading clinic was recently extended to children with other hematological diseases, including hemophilia and bone marrow failure. This has resulted in an opportunity for students with different diagnoses to teach their peers about their illnesses. Future goals include serving a larger number of children with SCD, providing longitudinal multiyear support, reducing barriers to attending the reading clinic, and exploring other methods of enhancing reading skills. In view of the many physical and neuropsychological challenges associated with SCD, interventions that result in improvement in reading skills offer an approach to significantly improve their achievement and quality of life.

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CONFLICT OF INTEREST

Winfred C. Wang received consultation fees from Agios and Novartis. The remaining authors had no conflicts of interest.

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