LOOKING DEEPER IN THE ASSESSMENT OF CHILDREN WITH SICKLE CELL DISEASE: A NEED TO CONSIDER LANGUAGE DEVELOPMENT AND LANGUAGE DISRUPTION ISSUES

Candice J. Adams-Mitchell, CCC-SLP.D
Department of Speech, Language, and Hearing Sciences, University of Florida, Gainesville, FL, USA

Fatima Jebahi, BSc (Hons)
Department of Communication Sciences and Disorders, East Carolina University, Greenville, NC, USA
Fulbright Association, Washington, DC, USA

— ABSTRACT —

Sickle cell disease (SCD) is a group of inherited red blood cell disorders that is caused by an abnormal type of hemoglobin (Hb, S) which causes red blood cells to become sickle-shaped and rigid. The burden of SCD results in chronic fatigue, neurological complications, frequent hospitalizations, pain, and adverse pharmaceutical side effects from medications used to manage SCD and illnesses associated with SCD. Individuals with SCD are at an increased of mortality and morbidity as a result of neurological infarcts, infections, acute chest syndrome, and vaso- occlusive pain crisis. Many children with SCD frequently experience lower school achievement and attainment relative to students without SCD. There are significant gaps in the literature regarding the strong association of SCD and speech and language development in children. We propose there is an inherent need for early therapeutic intervention for cognitive and language development and disruption issues along with academic support for the many children with SCD who experience frequent hospitalizations and subsequently school absences thus impacting their academic performance.

Keywords: Sickle cell disease; child language; cerebrovascular accidents, language development; cognitive-linguistic function

Introduction

Sickle cell disease (SCD) is a group of inherited red blood cell disorders that affects millions throughout the world. According to the Centers of Disease Control and Prevention, it is estimated that about 100,000 people in the United States, mainly Blacks or African Americans, suffer from SCD (Centers for Disease Control and Prevention, 2020). These data also indicate that 1 in 400 African American newborns in the United States are born with the condition. SCD is associated with an increased risk of early death (Maitra et al., 2017) and adverse clinical complications impacting a variety of organ systems (Miller et al., 2000). In turn, patients with SCD and their families report significantly diminished quality of life (Dale, Cochran, Roy, Jernigan, & Buchanan, 2011).

Sickle Cell Disease and Stroke

One of the most serious complications of SCD is stroke, which occurs in 11% of children younger than 20 years of age with the condition, making it 221 to 300 times more common in children with SCD compared to healthy children (Earley et al., 1998; Ohene-Frempong et al., 1998). Further, children with SCD and abnormal brain imaging findings due to strokes show greater occurrence of cognitive impairment (verbal intelligence and verbal comprehension) than those with normal imaging findings (Steen et al., 2003). The resulting cognitive impairments could be due to diffuse brain injury as well as a host of other factors such as socioeconomic status, drug and alcohol exposure, inadequate nutrition, untreated attention deficit and hyperactivity disorders (ADHD), along with chronic brain hypoxia (Steen et al., 2003).

There is also evidence that many children with SCD experience silent infarcts that are subsequently corroborated by imaging. Webb and Kwiatkowski (2013) found that 22% of children with SCD between 6 and 19 years of age experience silent infarcts. Unfortunately, even silent infarcts in children with SCD can leave many with significant neuropsychological deficits that appear to decline over time. Interestingly, Wang et al. (2001) found that school-aged children with SCD and silent infarcts exhibited lower scores in math and reading with their performances declining over time. Further, there is evidence that many children with SCD and no brain abnormalities can exhibit lower intelligence quotients (IQ) when compared to healthy controls (Kawadler, Clayden, Clark, & Kirkham, 2016). It is also important to note that there is a strong association between the presence of stroke and lower educational attainment in children with SCD (King, DeBaun, & White, 2008). At the same time, other biological, socioeconomic, and environmental factors associated with this population may also contribute to observed declines in IQ scores and overall reductions in educational attainment (Kawadler et al., 2016).

Sickle Cell Disease and its Comorbid Conditions

Along with greater risk of stroke and the neuropsychological consequences, asthma and acute chest syndrome (ACS) are comorbid conditions often seen in children with SCD. Children with SCD and asthma are at increased risk for developing ACS resulting in reduced lung function, and for some death (Knight-Madden, Barton-Gooden, Weaver, Reid, & Greenough, 2013). Common treatments for ACS and reduced lung function include corticosteroids and bronchodilators for the aggressive treatment and prophylactic management of asthma and ACS have known adverse effects on the central nervous system that contribute to cognitive performance issues. Research by Sharma, Hashmi, & Chakraborty (2020) offer evidence that bronchodilators increase excitatory responses which in turn result in issues that mirror attention deficit hyperactivity disorder (ADHD) symptoms. At the same time, corticosteroids are known to decrease serotonin levels which are also associated with ADHD and oppositional defiant disorder (ODD) (Saricoban et al., 2011). In summary, common medical therapies designed to improve quality of life can negatively influence cognitive performance.

Sickle Cell Disease and Cognition, Language, and Learning

Given the impact of medical therapies on neuropsychological functioning, there should also be concern regarding a range of other key developmental milestones and in particular language performance. Language performance is closely associated with neurological performance and commonly disrupted after neurological disorders such as stroke or other higher- level cortical disease processes. More specifically, speech, language, and learning disorders are common consequences of stroke in adults. The concern among children lies in studies that have shown that strokes occur in 25% to 60% of children with SCD (Balkaran et al., 1992; De Oliveira, Ciasca, & Moura-Ribeiro, 2008). Further concerns emerge from evidence that suggests cognitive decline can occur even in the absence of stroke. Consequently, one might hypothesize that language issues may also be present in the absence of stroke given their strong association.

Additionally, while language, and learning deficits have been largely documented in children with SCD, these same deficits are rarely described in significant detail to determine their long-term impact. For example, Hariman, Griffith, Hurtig, and Keehn (1991) noted in their study that their cohort of children with SCD and stroke presented with a variety of compli-

cations including severely impaired speech. However, specific speech and language diagnoses were not provided. Interestingly, De Oliveira et al. (2008) concluded that children with SCD and a history of stroke often present with acquired aphasia, learning difficulties, lack of initiative, and cognitive impairment all believed to derive from silent infarction prior to documented stroke onset. Similarly, Buchanan, James-Herry, & Osunkwo (2013) reported a case series of 5 children with SCD and a history of strokes where speech difficulties have been reported in case-descriptions, however not elaborated. Furthermore, specific deficits have been noted in various domains of language, including deficits in reading and writing skills (Sanders et al., 1997) as well as receptive and expressive oral language skills evident in difficulties following commands and formulating sentences respectively (Davis et al., 1997).

To date, little is known about how SCD specially disrupts speech and language development among children with SCD, many of whom are concurrently experiencing silent infarction even in the absence of a clear diagnosis of stroke. Yet there is some evidence that children with SCD exhibit language processing problem whether they are at low or high neurological risk (Schatz, Puffer, Sanchez, Stancil, & Roberts, 2009). The authors note that these issues are of major concern for children because language processing is linked to a range of language development skills. Of similar concern is the impact of SCD on speech and language skills in combination with the high frequency of missed school days due to SCD complications. In a large study in the UK, Dyson et al. (2010) found that children with SCD on average missed 8% of the school year or over 16 days. Over twelve percent of their sample missed 32 or more days. In a smaller study in the US, Schatz (2004) found that school-aged children missed on average 10% of the school year or 18.2 days each school year. They also noted that the significant number of missed days contributed to higher rates of grade retention relative to children with SCD. Overall, many children with SCD frequently experience lower school achievement and attainment relative to students without SCD (Swanson, Grosse, & Kulkarni, 2011). Given this likelihood of speech and language issues being present in children with SCD regardless of stroke and their relationship to school achievement, formalized approaches to assessment, monitoring, and management should be considered.

Conclusion

In conclusion, effective management of SCD requires knowledge and awareness of the disease itself but also commonly observed neurological implications that can negatively impact cognitive and language performance. While it is not known how the disease in combination with its consequences of stroke, and in some cases only silent infarction, disrupt or impact cognitive and language development and subsequently performance. Regardless, there is an inherent need for early therapeutic intervention for cognitive and language development and disruption issues along with academic support for the many children with sickle cell disease who experience frequent hospitalizations and subsequently school absences that impact their academic performances. These issues should be considered along with frequent chronic pain, fatigue, and adverse pharmacological side effects that impact cognitive and language performance. Beyond the traditional SCD team members, children with SCD could also benefit from early and ongoing consultations with speech-language pathologists given their expertise in cognitive, speech, and language development and knowledge of neurologically based conditions such as stroke that also have a negative impact cognitive, speech, and language performance.

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Contact Information:
Candice J. Adams-Mitchell, CCC-SLP.D
Email: c.adamsmitchell@phhp.ufl.edu