Sickle Cell Disease: Clinical Implications for Speech Language Pathologists

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Abstract
Individuals with Sickle Cell Disease (SCD) often exhibit a variety of comorbidities that have been described by professionals of many disciplines in different scientific journals, using varying terminology. This is an overview of possible disease maturated pathologies associated with SCD that fall under the Speech Language Pathologists’ (SLP) scope of practice.

Introduction
SCD is the most common autosomal recessive hereditary disorder recognized both nationally and internationally by the World Health Organization. SCD is characterized by misshapen red blood cells which cause sickle cell crisis. The most common crisis are vaso-occlusive and result in severe episodic pain and decreased oxygenation. Due to the encompassing nature of the vascular system these crisis subsequently affect multiple organ systems. Organs affected inherently include the brain which houses speech and language control centers. Areas of ASHA’s Big 9 that have been investigated within the SCD population include: expressive/receptive language, hearing, cognitive communication, and social communication.

While it is an SLP’s ethical responsibility to treat impairments in the aforementioned areas, they are also responsible for counseling and upholding ethical and professional integrity in others. Little research discusses ethical/professional matters of the chronically ill clinician.

Aims: The aim of this paper was to compile evidence that:
1. Described impairments associated with SCD that fell within the SLP’s scope of practice
2. Described the possible relationship between individuals with SCD and impaired practioning, the role of co-worker/colleague in ethical/professional dilemmas, signs/symptoms of decreased ethical/professional alretness, and preventative measures.

Methods
Four databases, Google Scholar and University of Delaware’s DELCAT Discovery was searched from inception to January 2019. Search terms included that of “Sickle Cell and __”. The blank included a variety of terms regarding clinical practice and ethics included varied combinations of the following: impaired practioning, ethical dilemmas, ethics, absenteeism, impaired clinician, impaired practice, sickle cell, and chronic disease.

Results
Neurological Manifestations:
There is a growing body of literature that attests to reoccurring findings of neurological manifestations noted and associated with individuals with SCD. Neurological deficits are one of the most devastating characteristics associated with SCD and depending on diagnosis, neurological changes may influence clinical presentations of the disease.12. Frequently investigated pathologies include but are not limited to:
- White Matter Hyperintensities - lesions to the white matter
- Lacunae- small cavity(whole) characterize as loss of tissue
- Atrophy- decrease in volume and mass of brain
- Vasculopathy - disease of blood vessels (e.g. small vessel)
- Ischemic/hemorrhagic strokes & silent cerebral infarcts (SCI)

Neurocognitive Manifestations:
Many of the neurological manifestations have implications cognitively. Neurocognitive and neuropsychological impairments that have been most frequently cited in those with SCD include:
- Intellectual quotient scores below normative data
- Executive functioning deficits
- Working Memory difficulties
- Processing difficulties
- Inhibitory & attentional issues

Investigators attribute these impairments to a variety of etiologies such as chronic hypoxia, anemia, poor pulmonary function, and SCIs.12,13. Individuals with impairments in the aforementioned areas often demonstrate difficulty learning and preforming well academically. Deficiencies in these areas have been observed to increase with age, therefore impacting QOL later in life.

Social Implications:
Social skills require competency in a variety of areas including theory of mind, attention, and working memory. Being that individuals with SCD are already susceptible to deficits in these areas preliminary research suggests individuals with SCD have difficulties with social skills and peer interaction secondary to executive functioning deficits. This phenomena has been documented among other brain injured populations (e.g. TBI, ADHD).

Language Implications:
Although, language impairments are frequently cited in other populations with central nervous system injuries (e.g. TBI, seizure disorders), the relationship between SCD and language impairments have not yet been clearly defined. Professionals may see varied standardized assessments, deficits in speech and language have been observed in semantic, lexical retrieval, syntactic and phonological/language processing abilities.

Results Cont.
Audiological Implications:
Both conductive hearing losses as well as sensorineural hearing losses have been found to occur more frequently in individuals with SCD compared to the general population. Causes are still poorly understood but are hypothesized to be associated with poor blood flow oxygenation to the auditory mechanisms. Depending on factors such as time of onset and severity, hearing losses have the potential to affect language learning, speech, behavior, cognition, social interactions and academics.

Practicing Ethically:
Up until this point, reasons why SLP’s may see individuals with SCD on their caseload has been discussed. However, SLP’s are more than just therapists, they are mentors, colleagues, employers, professors and etc. This section is dedicated to discussing the experience of the working healthcare professional, specifically the SLP with SCD. Due to the complex morbidities associated with SCD, there are possible implications regarding work/occupational QOL. Impaired Practioning is a term defined by the American Speech-Language-Hearing Association that occurs when a clinician’s professional and ethical practice are negatively impacted secondary to addiction, substance abuse, or health-related and/or mental health disorders.

Possible health-related implications related to SCD are:
- Physical inabilities
- Stress/ Burnout
- Presenteeism/Abseenteism
- Other

With this, the affected individual as well as peers, colleagues, mentors and employers should be able to identify signs and symptoms of decreased alertness of ethical/professional obligations. Signs/symptoms may be overt or implicit and highly variable. Regarding individuals with SCD one should understand and be aware of:
- Crisis Triggers and other physical signs of impairment
- Reduced energy, impaired concentration, depersonalization, deterioration of relationships, depressed quality of performance and etc.

To ensure professionalism, ethics and a healthy work atmosphere are preserved both affected individuals and associates should establish open communication and a situational protocol.

Clinical Implications/Conclusion
The primary purpose and focus of this paper was to evaluate SLP’s about the comorbidities associated with SCD that often go undetected secondary to a lack of awareness. It is important that SLP’s are knowledgeable about SCD so that they may advocate effectively and appropriately when warranted. Early identification and intervention is crucial for maintaining and improving cognitive, social, language, and hearing skills. Therefore, routine screenings, evaluations and neuroimaging are necessary to better understand deficits and facilitate improved QOL amongst individuals with SCD.

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References