The Role of Child Life Specialists in Transitioning Patients With Sickle Cell Disease From Pediatric Care to Adult Management

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THE ROLE OF CHILD LIFE SPECIALISTS IN TRANSITIONING PATIENTS WITH SICKLE CELL DISEASE FROM PEDIATRIC CARE TO ADULT MANAGEMENT

A Thesis

Submitted to the Graduate Faculty of the Louisiana State University and Agricultural and Mechanical College in partial fulfillment of the requirements for the degree of Master of Science

in

Child and Family Studies

by

Amanda Ochiltree
B.S., Louisiana State University, 2016
May 2022
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ABSTRACT

The transitional period when adolescents and young adults (AYA) no longer see pediatric specialists or pediatricians and transfer their care to adult health care providers is important for all but can be especially complex for AYAs with sickle cell disease (SCD). Disease-specific education, patient and family resources, and psychosocial care are important during the transitional period from adolescence to adulthood. Certified Child Life Specialists (CCLs) are trained to provide developmentally appropriate preparation and education, psychosocial support, encouragement of emotional expression, and the opportunity to form trusting relationships with the medical care team (Romito et al., 2021). CCLs work with the multidisciplinary team (e.g., doctor, social worker, nurse, other medical professionals) to provide care during the transition period. The findings from this study identified categories of child life interventions during the first three transition visits for patients with SCD. Introduction of self and services, educational assessment, completion of an "All About Me" form, and rapport building were child life interventions present during Transition Visit 1. Patient education, family planning, provision of handouts, and educational assessment were child life interventions present for Transition Visit 2. Discussion and review of pain plan, education related to non-pharmacological pain management, deep breathing demonstration, and guided imagery demonstration were child life interventions present during Transition Visit 3. The findings from this study can provide implications for CCLs working with patients who have SCD and face complex challenges associated with disease management and transitional care, and support the importance of child life documentation in the electronic medical record (EMR).
INTRODUCTION

Adolescents and young adults (AYA) face many challenges when transitioning from pediatric health care to adult management. This research study identified the interventions that Certified Child Life Specialists (CCLSs) provide for patients with sickle cell disease (SCD) transitioning from pediatric health care to adult management. CCLSs work with other members of the multidisciplinary team (e.g., doctor, social worker, nurse, other medical professionals) to provide care during the transition period. Bronfenbrenner’s ecological theory was used to provide a theoretical perspective for AYA with SCD transitioning into adult care.

The transitional period when AYAs no longer see pediatric specialists or pediatricians and transfer their care to adult health care providers is important for all AYA but can be an especially complex process for adolescents who have chronic health issues. Common chronic health issues among children and adolescents include SCD, asthma, diabetes, cerebral palsy, inflammatory bowel disease, congenital heart disease, and various other complex diagnoses (Chafe et al., 2019). It can be difficult for adolescent patients to have the desire to leave pediatric doctors, and it can be challenging to find health care providers and specialists to provide the appropriate care that is needed. In addition, there are often gaps in insurance coverage and lack of disease-specific education that affect transition readiness (Chafe et al., 2019).

Disease-specific education, patient and family resources, and psychosocial care are important during the transitional period from adolescence to adulthood. CCLSs are trained to provide developmentally appropriate preparation and education, psychosocial support, encouragement of emotional expression, and the opportunity to form trusting relationships with the medical care team (Romito et al., 2021). Although literature is limited, most literature
identifies child life services in hospitals and outpatient clinics as providing psychosocial preparation, education related to diagnosis and treatment, pain management and coping strategies, therapeutic play, and family support (Romito et al., 2021). Anecdotally, in practice, CCLSs provide comprehensive care for AYAs with SCD in pediatric hematology and oncology (Johns Hopkins All Children’s, 2021; Northwell Health Foundation, 2021; St. Jude Children’s Research Hospital, 2021; UCSF Benioff Children’s Hospital, 2021). There is currently little information and a gap in the research related to CCLSs working in hematology and oncology clinics with transition programs for patients with SCD. The American Academy of Pediatrics (2021) recommends additional research to evaluate the effects of child life services on patient care outcomes. Before we can do that, we first must identify what interventions CCLSs are offering to patients with SCD.
REVIEW OF LITERATURE

A review of literature was conducted to assess common stressors for AYA with SCD and related child life interventions. First, SCD will be discussed including disease-specific symptoms and management. Then, the health care transition process will be explained. Finally, a general overview of child life services will be provided to assist in defining the work of a CCLS in transitional care for AYAs with SCD. The literature will be used to establish the types of possible child life interventions that are offered to AYA patients with SCD and identify child life interventions that could be offered to AYA patients with SCD.

Sickle Cell Disease

Physiological Symptoms

SCD is an inherited red blood cell disorder that affects approximately 100,000 individuals in the United States (Centers for Disease Control and Prevention [CDC], 2020). Individuals that have SCD do not have normal hemoglobin that contains healthy red blood cells; instead, their red blood cells are rigid and sickle-shaped (CDC, 2020). Healthy red blood cells are small and flexible, allowing the red blood cells to fit through blood vessels throughout the body (CDC, 2020). Unlike healthy red blood cells, the irregular, crescent shape causes sickled cells to get stuck and block blood flow to organs throughout the body (CDC, 2020). In result, individuals with SCD can experience life-threatening complications and health concerns (e.g., pain crisis, anemia, infection, acute chest syndrome, stroke, organ damage, or vision loss; CDC, 2020; Hassell et al., 2016).

SCD can impact the reproductive health of both men and women. Studies show that some reproductive issues are caused by chronic medical therapy and treatment of SCD (Smith-Whitley, 2014). Reproductive issues can include delayed puberty for both sexes by one-to-two
years, priapism (i.e., unwanted painful erection that lasts for several hours) and/or sperm abnormalities in men, and hormonal fluctuations in women (Smith-Whitley, 2014). In men, priapism may cause hypogonadism, sperm abnormalities, and erectile dysfunction (Smith-Whitley, 2014). Smith-Whitley (2014) explained that more research is needed to understand the impact that medical therapies (i.e., chronic transfusion, hydroxyurea, and hematopoietic stem cell transplantation) have on fertility and reproductive issues for people with SCD.

**Diagnosis and Screening**

SCD is genetically inherited and is most common among racial and ethnic minorities in the United States (Hassell, 2016). Unlike other chronic pediatric diseases, SCD is highly prevalent in African Americans in the United States (Barakat et al., 2006). SCD also can affect Hispanic Americans, and people of Middle Eastern, Asian, Indian, and Mediterranean descent (Yusuf et al., 2011).

There are several forms of SCD that vary in severity and are characterized by different genotypes: sickle cell anemia, sickle-hemoglobin C disease, two types of sickle beta-thalassemia, and a few rare types of SCD that are caused by an abnormal type of hemoglobin (CDC, 2020; Yusuf et al., 2011). Additionally, individuals that inherit a sickle cell gene from one parent and a non-sickle cell gene from the other parent have sickle cell trait (SCT). Individuals with SCT do not have symptoms of SCD but can pass SCD or SCT to their children (CDC, 2020). Approximately one in every 13 Black or African American babies is born with SCT (CDC, 2020).

The most serious form of SCD is sickle cell anemia in which individuals inherit hemoglobin S genes from both parents (CDC, 2020). A milder form of SCD is sickle-hemoglobin C (CDC, 2020). Individuals with sickle-hemoglobin C inherit the hemoglobin beta S
gene from one parent and the hemoglobin C gene from the other parent (CDC, 2020). Sickle beta plus thalassemia and sickle beta zero thalassemia disease are two types of SCD that are distinguished by the amount of hemoglobin produced (CDC, 2020). A person with sickle beta thalassemia inherits one sickle cell gene from one parent and one beta thalassemia gene from the other parent (CDC, 2020). Sickle beta plus thalassemia is a milder form of SCD compared to sickle beta zero thalassemia (CDC, 2020).

SCD can be determined by prenatal testing but is usually diagnosed using the universal newborn screening at the time of birth (Yusuf et al., 2011). In 2006, the United States began requiring universal newborn screening for SCD, which has allowed the opportunity for parents, caregivers, and health care providers to be informed of the diagnosis (Benson & Therrell, 2010; Manley, 1984).

Many studies have been done to examine the attitudes and beliefs related to prenatal genetic testing for SCD. In the future, noninvasive prenatal genetic testing and prenatal genetic counseling will be more readily available to provide individuals with prenatal diagnosis and resources to prepare for a child with SCD (Hill et al., 2014). Although the universal newborn screening is required, the use of genetic counseling following a positive newborn screening is low in the United States (Stevens et al., 2019).

Genetic counseling allows for the opportunity to have disease-specific care and additional resources to equip caregivers with knowledge and understanding of SCD. Unfortunately, there is not a standard approach to follow-up programs and communication related to a positive diagnosis of SCD or SCT at the time of the newborn screening (Stevens et al., 2019). Patients and families experience challenges due to lack of follow-up support and limited communication from health care providers. Benson and Therrell (2010) argued that the universal newborn
screening programs should include better short- and long-term care, attention to carrier detection, and education (e.g., SCT identification). SCD symptoms do not begin until approximately four months of age; therefore, if prenatal testing is not completed, the newborn screening is the only initial indicator of the SCD diagnosis (Quinn, 2013).

**Risk and Resilience Factors**

SCD can cause many health risks and complications but can also have a life-long psychological impact. Depending on the type of SCD, daily management and treatment can differ, but interference with daily functioning (e.g., school, extracurricular activities, and normative peer interactions) is often the result of SCD symptoms and complications (Barakat et al., 2006). Psychosocial stress can enhance physiological symptoms (e.g., pain and the inability to participate in social and physical activities; Shah et al., 2020). Shah et al. (2020) found that mental stress caused a decrease in blood flow, thus triggering pain crisis for patients with SCD. Psychosocial care should be taken into consideration to better understand how children and adolescents adapt to living with the chronic disease.

Most individuals with SCD in the U.S. are from underserved African American and Hispanic communities that depend on health resources (i.e., public insurance and healthcare programs; Hassell, 2016). Findings from Hassell’s (2016) study addressing areas of SCD disparities concluded that family and friends of individuals with SCD are relied upon as trusted resources for health and genetic information. Furthermore, Long et al. (2011) discovered that barriers to disease-specific education and awareness were a result of fear, lack of communication, and sociocultural components (e.g., African American culture and mistrust of healthcare professionals). Nine percent of AYA patients with SCD reported concern associated with mistrust from doctors regarding pain medication (Sobota et al., 2014).
Furthermore, race and culture are important considerations when understanding risk and resilience factors for individuals with SCD (Barakat et al., 2006). In Kusyk et al.'s (2013) study related to prenatal and neonatal genetic testing in postpartum African American women, 51% of postpartum women remembered their discussion with a pediatric provider after their infant had the newborn screening. Furthermore, 25% of postpartum African American women did not know their hemoglobinopathy status (Kusyk et al., 2013). Long et al. (2011) described the lack of awareness of being a carrier of SCT was also a barrier to providing parents with education and identification of SCT. Additionally, 42% of participants in Long et al. (2011) study knew their SCT status.

In 1972, Congress passed the National Sickle Cell Anemia Control Act, which resulted in changes in legislation and funding for SCD. Greater awareness and funding resulted in better education, counseling, and treatment of SCD (Manley, 1984). Furthermore, SCD screening clinics have been established under the National Sickle Cell Anemia Control Act, providing education to individuals that previously did not have the disease-specific resources (Manley, 1984).

**Treatment and Maintenance**

SCD treatment and maintenance differ depending on the severity of the disease (CDC, 2020). Signs and symptoms can start as early as five months of age, therefore identification of SCD allows for comprehensive care for infants. Prior to five months of age, hemoglobin F (i.e., fetal hemoglobin) prevents the red blood cells from sickling (CDC, 2020). Treatment includes penicillin prophylaxis (antibiotics), pneumococcal immunization, hydroxyurea therapy, transcranial doppler screening for stroke prevention, and opioids used for pain management (Martin et al., 2018). Because SCD can worsen over time, some disease-specific complications
can be reduced by compliance with comprehensive treatment and maintenance, but with most treatments, individuals can still have extreme pain episodes, organ damage, and reduced life expectancy (CDC, 2020).

Hydroxyurea is a medication used to decrease the complications of SCD by making red blood cells bigger. By increasing the amount of hemoglobin F and hemoglobin, the red blood cells become bigger, appearing round and more flexible, therefore reducing the likelihood of the cells becoming sickled (Agrawal et al., 2014). Hydroxyurea reduces pain crisis by 50% (Agrawal et al., 2014). Additionally, hydroxyurea is effective in reducing the amount of blood transfusions needed and number of acute chest syndrome episodes (Agrawal et al., 2014).

Although the American Society of Hematology (2020) established professional guidelines that emphasize the importance of antepartum care, perinatal care, and acute and chronic pain, there is still a lack of awareness of disease-specific characteristics (Brandow et al., 2020). Martin et al. (2018) found that parents were highly satisfied and had better adherence to medications and treatment plans after receiving resources from the comprehensive care clinic. When resources are provided to parents of children with SCD and caregivers are supported, children and adolescents may better adapt to disease-specific stressors (Martin et al., 2018).

Advanced medical treatment and maintenance has reduced mortality rates and increased medical intervention for children with SCD (CDC, 2020). The Cure Sickle Cell Initiative was started in 2018 to support SCD clinical research associated with new advancements in gene therapy (U.S. Department of Health and Human Services, n.d.). Ashorobi and Bhatt (2021) found promising results using gene therapy to remove stem cells from bone marrow and introduce a therapeutic gene to produce anti-sickling cells. Bone marrow transplant (BMT) in SCD has many risks and is only considered an option for individuals with severe disease.
Future clinical research and funding supporting SCD will aid in the development of new treatment and a potential cure for SCD.

**Psychological Repercussions**

AYA patients with SCD may experience psychological repercussions due to their chronic illness (Hicks & Davitt, 2009). There are mental health risks (e.g., anxiety, depression), socioemotional challenges, and unique issues experienced by AYAs with SCD (Anie, 2005). Pain experienced by children and AYAs with SCD can impact quality of life, including socialization with others, and can lead to disruption in routine (e.g., absence from school, inability to participate in physical activity; Howard et al., 2009). AYAs can experience increased difficulty with coping and adaptation to disease-specific stressors (Hicks & Davitt, 2009). Furthermore, inappropriate pain management and coping strategies and frequent emergency department visits have been identified in children and adults with SCD (Anie, 2005; Yusuf et al., 2010).

**Health Care Transition**

The health care transition process for AYAs with special health care needs is a popular topic among youth, families, and health care providers. Successful self-management includes understanding disease-specific symptoms and treatment, medication compliance, adapting to the challenges and barriers related to the complexity of the disease, and engaging in active communication with the health care team to improve their overall quality of life (Ahmadi et al., 2015). The transition period is generally expected between ages 18 to 21 years (Goralski et al., 2017). Research shows that the expectation for transition in care occurs based on age, not maturity or readiness (Feinstein et al., 2017; Reiss et al., 2005).
To prepare for self-management to meet their unique health care needs, patients with SCD require education about their medical diagnosis, plan of care, and specific medical concepts. Ferris et al.’s (2012) health transition scale has been used over time to repeatedly assess the patient as their knowledge, independence, and mastery of self-management evolves. Due to the complexity of health care transition based on the type of illness, resources, self-management and knowledge, the transition scale allowed health care providers to have a reliable and valid measure of skill mastery and areas of importance for transition readiness (Ferris et al., 2012). Because the health transition scale incorporates an interview, the healthcare provider can give the patient feedback and reassurance throughout the transition period (Ferris et al., 2012).

Additionally, Goralski et al. (2017) described the Transition Readiness Assessment Questionnaire as another validated tool for cystic fibrosis patients to adequately measure and assess the skills needed for an effective transition (e.g., self-advocacy and self-management). Research shows that self-advocacy and self-management are expected for transition readiness, but age is also an expectation for the transition (Goralski et al., 2017). Little is known about how physicians approach lack of readiness at the expected age of transition. When AYAs do not have independence and mastery of self-management at the time of transition, they can experience gaps in health care, deterioration of health, and decrease in quality of life (Feinstein et al., 2017).

Sobota et al.’s (2014) study on patients with SCD ages 18-22 seen in a SCD transition program over a two-year period reported understanding of SCD, knowledge of medications, and comprehension of genetic inheritance of SCD. Additionally, in the transition assessment tool, the majority of AYAs with SCD reported having plans for their future, good social support, and knowledge of how to refill prescriptions (Sobota et al., 2014). About 64% of patients understood
factors associated with health insurance (i.e., age and health insurance card information; Sobota et al., 2014).

**Health Care Transition Challenges and Barriers**

There are many complex challenges and barriers that individuals face during the time of adolescence and young adulthood in addition to having to navigate the transition to adult health care providers. For example, lack of preparedness and separation from their pediatric provider are major barriers to successful transitions (Bemrich-Stolz et al., 2015; Feinstein et al., 2017). In addition, inadequate insurance coverage and lack of specialized adult health care providers have been challenging for patients and families. Specifically, patients with SCD had concerns about medical cost and treatment with adult health care providers (Telfair et al., 1994).

Disease-specific transitioning needs to be considered when discussing the transition for adolescents. Zablotsky et al. (2020) conducted a study to explore health care transition and planning specifically related to youth with autism spectrum disorder and other mental, behavioral, and developmental disorders. Youth with autism spectrum disorder were less likely to have completed health care transition items (e.g., actively working with their doctor, scheduling independent doctor visits, and having a parent who knows the insurance plan for adulthood) compared to youth with other disorders (e.g. mental, behavioral, and developmental; Zablotsky et al., 2020).

Age of the individual should be considered when understanding the transitional process and possible outcomes. Feinstein et al. (2017) assessed cystic fibrosis (CF) patients and the changes in outcomes for patients as health care services have evolved. CF patients' survival rates have increased, and they now live to be older than 18 years of age. Moreover, because of the improved outcome of CF patients and resources, the Cystic Fibrosis Foundation mandated that
CF centers develop specific programming to transition patients to adult management by 21 years old (Feinstein et al., 2017). This is a specific age requirement and expectation for all patients. After McLaughlin et al. (2008) studied CF centers and found that the transition period was beginning at 17 years and was not effective in providing the proper medical history and addressing important topics (e.g., smoking, diet, substance abuse), the transitional period was changed to begin much younger (e.g., 12-13 years) with age-specific goals for the patient. Goralski et al. (2017) found that patients who discuss a structured, planned transition at an early age have increased satisfaction with their pediatric center and a positive perception of their health and prognosis.

There is not an organized system of care for SCD like there is for CF, hemophilia, or other chronic illnesses (Hassell, 2016). Fenton et al. (2015) explored the relationship between health care transition-readiness and disease-specific characteristics, psychosocial factors, and health care outcomes among adolescents with chronic kidney disease. Transition readiness was based on fewer visits to the emergency room and better medication adherence, but family cohesion (e.g., an integrative and supportive family system) was the only significant psychosocial predictor of transition readiness (Fenton et al., 2015).

Furthermore, studies have shown that barriers and challenges for transitioning AYAs into adult health care settings vary based on the patient and their chronic health condition. In the first study exploring the transition period among individuals with SCD, Telfair et al. (1994) found that patients were concerned about medical cost and treatment from adult health care providers. For congenital heart disease, the American Heart Association (2011) stated that the ideal transition program is a goal but may not be achievable based on the current state of healthcare. There is not a universal approach to an adequate transition process, but family cohesion remains
an important factor in transitioning from pediatric care to adult management. In addition, strong relationships, developmentally appropriate goals, shifting the interaction to be patient-led, and preparing parents are all important factors in a successful transition process (Fenton et al., 2015).

**The Child Life Profession**

Chronic illness and hospitalization can be detrimental to children's development as children experience trauma, stress, disruption in routine, and regression (Turner, 2009). CCLSs are healthcare professionals trained to provide psychosocial care for children and families to aid in coping and adjustment during health care experiences (Romito et al., 2021). The psychosocial care of children within hospitals is essential to promote patient- and family-centered care, healthy emotional expression, and children’s abilities to explore and learn through their experiences and environment (Koller, 2008; Thompson, 2009).

CCLSs work with pediatric patients in various settings (e.g., pediatric inpatient units, outpatient clinics, emergency departments, hospice and palliative care settings, and chronic care centers) to provide ongoing support and education (Romito et al., 2021). Child life presence and support ensures that the unique developmental and cultural needs (i.e., backgrounds, experiences, and beliefs) of children and families are appropriately met (Boles et al., 2020). Developmental assessments, appropriate interventions, and cultural responsiveness in child life practice aids in building rapport with patients and families to reduce the stressors associated with chronic illness and hospitalization (Thompson, 2009).

Based on their assessment of the child’s cognitive and socio-emotional development, CCLSs implement individualized interventions to reduce stress and aid in coping with stressful medical encounters and adaptation to both acute and chronic illnesses (Thompson, 2009). In practice, CCLSs also assist in managing pediatric pain and distress by using non-
pharmacological pain management and creating opportunities for self-expression. Specific interventions include therapeutic and medical play, diagnosis-specific education, procedural preparation and distraction, and family and sibling support (Beickert & Mora, 2017; Romito et al., 2021).

**Patient- and Family-Centered Care**

Patient- and family-centered care (PFCC) is an approach to health care adopted by doctors, nurses, CCLSs, and other members of the interdisciplinary team that recognizes and honors the following principles: dignity and respect for the child and family; information sharing; participation; and collaboration (Johnson & Abraham, 2012). In the late 20th century, PFCC became an important aspect of health care as the awareness of meeting the psychosocial and developmental needs of children increased (American Academy of Pediatrics, 2003). The American Academy of Pediatrics (2021) considers child life services to be a quality guiding principle of PFCC and an important contribution to health care experiences. Child life presence and support can be used to mitigate the challenges and barriers experienced by patients and families, especially for adolescents and emerging adults during the transition process from pediatric care to adult management.

Parent-child relationships, communication, and behavior differ across cultures (Linton & Green, 2019). Culture is a central process of human development, and this is especially seen in children's play and behavior across cultures (Velez-Agosto et al., 2017). Understanding communication differences, religious and spiritual beliefs, complex family dynamics, and perceptions associated with health care provides insight on the specific challenges and barriers faced by children and families (Linton & Green, 2019). Although poverty and other economic inequalities have decreased, disparities among Black and Hispanic families continue to exist that
can result in limited access to basic healthcare resources (Linton & Green, 2019). CCLSs provision of PFCC, including cultural responsiveness, results in enhanced communication, increased patient and family engagement, and better patient outcomes (Bell et al., 2009). Feinstein et al. (2017) found that the difference in an adult-centered model has led to many challenges for adolescents and families who are accustomed to the supportive PFCC seen in the pediatric health care setting.

**History of the Child Life Profession**

The child life profession originated in the 1920s, when CCLSs were known as "play ladies" (Beickert & Mora, 2017). C.S. Mott Children’s Hospital in Michigan and Babies and Children’s Hospital of New York were a few of the first health care organizations to create programs that used play to address the fear and anxiety related to hospitalization (Beickert & Mora, 2017). The 1970s were a time of increasing growth for child life programs throughout North America (Association of Child Life Professionals, n.d.). In 1997, the mission, values, and vision of child life services were published, including the operating principles that are followed today (Association of Child Life Professionals, n.d.). The Child Life Council helped to create the certification process for CCLSs in 1998 (Association of Child Life Professionals, n.d.). In 2016, the Child Life Council changed their name to the Association of Child Life Professionals (Association of Child Life Professionals, n.d.).

**Child Life Specialist Requirements**

CCLSs must have earned a bachelor’s degree and completed ten courses related to child life, including a child life course taught by a CCLS, a minimum of two child development courses that cover birth to age 18, a family systems course, a play course, a loss and bereavement/death and dying course, a research course, and three additional courses related to
child life content (Association of Child Life Professionals, n.d). In addition to the specific course requirements, child life students must complete clinical training (i.e., 600-hour internship) under the supervision of a CCLS (Association of Child Life Professionals, n.d). Additionally, after completing an eligibility assessment, a comprehensive certification exam is required to become a CCLS (Association of Child Life Professionals, n.d). The ACLP has established the requirements to ensure that CCLSs are equipped with the knowledge and expertise to address the psychosocial needs of patients and families (Association of Child Life Professionals, n.d).

**Psychological Preparation and Support**

CCLSs provide psychological preparation and support for medical encounters to reduce stress and aid in coping for hospitalized children (Koller, 2008; Sisk, 2016; Thompson, 2009). CCLSs provide developmentally appropriate verbal preparation and incorporate medical materials and teaching dolls to prepare patients for procedures and imaging (Koller, 2008). CCLSs use honest and concrete words to explain medical procedures in a way that the patient can understand to address potential misconceptions the patient may have, depending on the patient’s developmental level (Koller, 2008). Providing the patient with sensory information and allowing the patient to manipulate the medical materials allows the patient to enhance communication and knowledge about the medical encounter (Goldberger et al., 2009; Koller, 2008).

**Diagnosis teaching and education**

In addition to providing psychological preparation and support, CCLSs also provide developmentally appropriate education at diagnosis of a chronic or acute illness (Goldberger et al., 2009). Developmentally appropriate communication related to illness can empower the patient to feel a sense of control and adapt to living with a chronic or acute illness (Hicks &
Davitt, 2009). CCLSs in hematology and oncology clinics utilize their knowledge and skillset to provide developmentally appropriate preparation and education for new hematology and oncology diagnoses, invasive procedures (e.g., surgeries, port access), therapy (e.g., chemotherapy, radiation, or blood transfusions) and imaging (e.g., magnetic resonance imaging [MRI], computed tomography [CT], positron emission tomography [PET] scan, or transcranial doppler ultrasound [TCD]). In a 2012 study, 71 children with SCD who received preparation and support prior to having an MRI were more likely to complete a clinically interpretable (clear) MRI without sedation and anesthesia (Cejda et al., 2012).

In addition, CCLSs incorporate therapeutic and medical play into their interventions to promote self-expression and allow patients the opportunity to manipulate medical materials, gain understanding, and address potential fears and misconceptions associated with their medical encounters and chronic diagnosis (Goldberger et al., 2009; Hicks & Davitt, 2009). CCLSs in hematology and oncology clinics consider socio-cultural context when facilitating legacy building activities and support at end of life (Sisk, 2016). Based on a CCLS’s assessment, in practice, appropriate patient and family resources are offered to optimize patient and family engagement and generate positive coping outcomes.

**Non-pharmacological pain management**

Pain experienced by children and adolescents can be complex and difficult to assess and manage (Srouji et al., 2010). Physiological responses, behavior, and self-report are used to assess pain levels (Srouji et al., 2010). The American Society of Hematology (2020) recommends non-pharmacological management (e.g., massage, yoga, virtual reality [VR], and guided imagery) for acute pain associated with SCD. CCLSs utilize developmental assessments to incorporate appropriate non-pharmacological pain management techniques to mitigate pain (Srouji et al.,
2010; Westbrooks, 2021). In practice, CCLSs use distraction techniques, guided imagery, VR, pet therapy, and essential oils in their interventions for SCD patients during heightened pain experiences (Westbrooks, 2021). Williams and Tanabe (2016) found that 12 of 28 studies on non-pharmacological pain management reported that non-pharmacological techniques were successful in reducing pain for patients with SCD. Additional research is needed to assess the efficacy of non-pharmacological interventions (Williams & Tanabe, 2016).

**Child Life Specialists' Role in the Sickle Cell Transition Clinic**

Based on the current literature, CCLSs assist in identifying and meeting potential needs of patients and families during the transitional period of adolescence to adulthood (Boles et al., 2020; Clayton-Jones et al., 2021). The period of adolescence and emerging adulthood is a stressful stage in a young adult's life that can result in difficulty coping, poor health outcomes, sudden onset of changes, and difficulties with access to appropriate care (Goralksi et al., 2017).

Clayton-Jones et al. (2021) found that several social and psychological factors contribute to the health care transition process for AYAs with SCD. In Clayton-Jones et al.'s (2021) qualitative study, AYAs expressed the need for support (e.g., peer support and community support) and assistance identifying realistic goals for their future (e.g., education and family planning). CCLSs are trained to assess AYAs’ understanding of their medical experience and provide individualized interventions to aid in the successful transition to adult healthcare management (Hicks & Davitt, 2009). Additionally, AYAs with SCD expressed that they felt a stigma related to their disease (Clayton-Jones et al., 2021). For example, they did not feel like active participants in their care (i.e., medication management and medical decisions were often predetermined) and at times felt isolated from peers (Clayton-Jones et al., 2021). CCLSs recognize potential cognitive and socio-emotional challenges and barriers faced by patients and
families (Boles et al., 2020). Furthermore, AYAs with SCD value support from their health care professionals (Clayton-Jones et al., 2021). Child life presence and support can be used to educate AYAs about disease-specific stressors, and provide opportunities for self-expression, socialization, and resources that can assist patients and families during stressful experiences (Sisk, 2016).

CCLSs’ assessments include child variables (e.g., age, developmental level, temperament), family variables (e.g., parental anxiety and distress, family characteristics, socioeconomic status, parental presence and involvement), illness variables (e.g., chronic, acute, length of hospitalization), and medical experiences (e.g., invasive procedures, previous medical encounters) that contribute to the patient’s and family’s coping and adjustment (Koller, 2008). Child life assessments are ongoing and continuous assessments that can change over time. Child life assessments include information from members of the interdisciplinary team (e.g., doctors, nurses, social workers) and family (Hollen & Skinner, 2009). CCLSs also consider culture, language, and social support in their psychosocial assessments to determine potential stressors that the patient and family may experience (Hollen & Skinner, 2009).

**Theoretical Perspective**

Bronfenbrenner’s ecological theory explains how an individual is shaped by levels of external influence (Bronfenbrenner, 1977). The process-context model provides a structure for assessing the impact of the external environment on family processes (Bronfenbrenner, 1994). Bronfenbrenner’s (1977) ecological theory includes the child developing within a complex system of relationships influenced by multiple levels of the environment: the microsystem, mesosystem, exosystem, macrosystem, and chronosystem. The environment acts as a series of interrelated structures that form the overall system (Bronfenbrenner, 1977).
The microsystem is the inner most part of the ecological model. The microsystem includes the developing individual’s immediate environment (e.g., family, peers, health services, religion). The SCD transition clinic would be considered part of the patient’s microsystem (Griffin et al., 2013). The mesosystem accounts for interactions within all ecological systems (e.g., microsystem and exosystem; Bronfenbrenner, 1977). For example, a parent or caregiver’s interactions with members of the medical team may influence the patient with SCD (Griffin et al., 2013). Additionally, the exosystem includes settings or social structures that can influence the individual, but do not require active participation and involvement by the individual (e.g., extended family, community services, or the parent's workplace; Bronfenbrenner, 1977). For a patient with SCD, a parent’s experience in the health care environment and community services may have an impact on the patient with SCD (Griffin et al., 2013).

The macrosystem influences the developing individual through beliefs, attitudes, and values within their culture and society (Bronfenbrenner, 1977). This does not include people or places, but instead includes the developing individual’s socioeconomic status, ethnicity, laws, and customs (Bronfenbrenner, 1977). The macrosystem explains how an individual is influenced by the relationship between religion, race, and class (Bronfenbrenner, 1977). AYAs who are transitioning from pediatric health care to adult management are influenced by cultural elements that can affect self-exploration and developmental processes (Griffin et al., 2013). For AYAs with SCD, socioeconomic status and cultural ideologies can impact the transitional process (i.e., access to transportation, distrust of health care providers).

The chronosystem is the outermost system within the ecological model (Bronfenbrenner, 1977). The chronosystem includes timing of transitions and environmental changes that influence an individual’s life (i.e., pandemic, new diagnosis, natural or human-made disaster).
The impact of the COVID-19 pandemic (i.e., quarantine, virtual health care visits) and timing of transition may have an impact on AYAs transitioning to adult management. Bronfenbrenner’s ecological theory can be used by CCLSs to understand the patient’s family system and make developmental assessments based on patient and family interactions and the health care environment (Griffin et al., 2013; Turner, 2009).

**The Present Study**

The purpose of this retrospective chart review was to analyze medical notes pertaining to SCD and child life services and identify the child life interventions provided for AYA patients during the first three transition visits with the healthcare team by the CCLS in a SCD transition clinic. This retrospective study explored the role of CCLSs working with patients aged 14 to 19 with SCD in a transition clinic within an outpatient pediatric hematology and oncology clinic. By completing a retrospective chart review in the electronic medical record (EMR), child life interventions will provide a comprehensive understanding of CCLSs’ role when working with patients with SCD transitioning from pediatric care to adult management. The research question guiding this study is: What interventions do CCLSs provide to AYA patients with SCD in transition clinic? The research question was used to guide the data collection and analysis for three points in time (e.g., Transition Visit 1, Transition Visit 2, Transition Visit 3).
METHOD

Research Design

The present study is a retrospective chart review used to understand what a CCLS’s role is when working with AYA patients with SCD during the health care transition process. There is little known about CCLSS’s roles working with AYAs during the health care transition process, and the feasibility of collecting data in a live healthcare environment can be challenging. Therefore, a retrospective chart review, which consisted of prerecorded data within the EMR, was collected and analyzed to answer the research question. Hospital and child life documentation standards can differ among hospitals but there are perceived benefits of charting for CCLSS (e.g., communication, assessment, productivity, accountability; Hollen & Skinner, 2009). CCLSS provide EMR documentation detailing their assessment, intervention, and perceived outcome (Hollen & Skinner, 2009; Parish & Johnson, 1987). The retrospective chart review research design allowed for existing data to be used and analyzed, to not impede on the live clinical environment, and to further the existing knowledge on the interventions provided by CCLSSs for AYA patients with SCD undergoing the healthcare transition process.

Sample

Total population sampling was used for this research study. Total population sampling is a type of purposive sampling used to study a subgroup within a larger population and is typically used when a small number of cases exist (Etikan et al., 2016). The present sample included AYA patients ($n = 18$) with SCD seen in a pediatric hematology and oncology outpatient clinic in the southeastern United States. The mean age for Transition Visit 1 was 16.02 years ($SD = 0.56$, $R= 15.02$ to 16.11 years). The mean age for Transition Visit 2 was 16.40 years ($SD = 0.40$ years, $R= 15.10$ to 17.01 years). The mean age for Transition Visit 3 was 17.02 years ($SD = 0.57$, $R= 16.02$ to 17.11 years).
16.01 to 17.10 years). Participants were Black (100%) and non-Hispanic or Latino/a (100%). Seven patients were female (39%) and 11 patients were male (61%).

All SCD transition patients scheduled for transition clinic appointments during the months of June 2019, July 2019, and August 2019 were included in the sample. For this sample, up until the date of IRB approval, all SCD transition clinic child life documentation in each patient’s EMR was collected as data and used for this study. This time period was selected because patients were likely to be out of school in the summer months and more easily able to attend scheduled appointments. In addition, there was known child life presence in the transition clinic in 2019 at the study site, prior to the COVID-19 pandemic.

**Procedure**

The retrospective chart review was approved by the Institutional Review Board at Franciscan Missionaries of Our Lady Health System/FranU (Appendix A) and a reliance agreement between this institution and Louisiana State University’s IRB was obtained and approved (Appendix B). After approval from both IRBs, a search in the EMR was done using month and date to identify scheduled transition clinic days in the months of June 2019, July 2019, and August 2019. Then, patients with SCD scheduled for a transition visit were identified by their appointment on each transition clinic day. A list of patients that met inclusion criteria was created and their medical records were accessed. When reviewing each patient’s medical record, a search for child life notes was completed. Then, sickle cell transition child life notes were accessed, de-identified (i.e., removal of the patient’s and CCLS’s names and any personally identifiable information) and labeled by participant number. The sickle cell transition child life notes were then copied into a document, saved as a PDF, and stored on the hospital’s network folder per IRB protocol.
Measures

CCLSs document child life interventions for each patient’s medical encounter. In the EMR, the child life chart note included a narrative, describing the child life services provided for the patient and their family members during the medical encounter. See Appendix C-E for sample child life transition visit documentation templates. Three visits for each participant were documented, thus providing three narratives for each participant. Additionally, the date of transition visit, age, date of birth, gender, race, and ethnicity were collected from the EMR. Date of birth was converted to age, and calculated dates of appointments were converted into time (i.e., years, months, and days). Therefore, the participants’ date of birth was never documented on stored data files or documents.

Data Analysis

The process of analyzing the qualitative data from the EMR included content coding and thematic analysis. The process of analyzing data was inductive by categorizing the CCLSs’ documentation with patterns and themes (Creswell, 2018). The chart notes were separated by visit type and divided into categories based on the clinic visit number (i.e., Transition Visit 1, Transition Visit 2, Transition Visit 3). Then, separately, without discussion, the author and a CCLS read each patient’s chart note and discovered themes within the data. There was approximately one paragraph of narrative for each visit note. Each theme was categorized based on existence of a word or phrase and coded if the words explicitly stated the concept, even if the text appeared in different forms. Then, the author and CCLS met to discuss the themes. During this discussion, some themes were combined, revised, or removed completely. Mutually agreed upon themes were discussed and defined. Then, separately and without discussion the texts were coded again by the same coders. This process was repeated an additional time to determine the
final categories, for a total of three times. Then, intraclass correlation coefficients (ICC) were used to interpret the reliability of themes among coders using SPSS 27, a statistical software program.
RESULTS

Child Life Intervention During SCD Transition Visit 1

To answer the research question at time 1, 18 EMRs were accessed, and 13 EMRs (n = 13) were found to have CCLS chart notes for Transition Visit 1. There were five missing chart notes. Results of the content coding and thematic analysis by the two coders indicated four categories of CCLS interventions during Transition Visit 1 for patients with SCD: (1) introduction of self and services, (2) educational assessment, (3) completion of an "All About Me" form, and (4) rapport building.

During Transition Visit 1 (i.e., the initial transition visit), the CCLS introduced themself and provided the patient with an introduction of child life services, which included the role of a CCLS, how the CCLS can best support the patient in the transition process, and education related to the program and child life role. For example, the CCLS stated, "CCLS met with pt and his mother to introduce self and services during transition clinic." Additionally, the CCLS documented, "CCLS explained role of child life during transition visits for support and education." The CCLS documented introduction of self and services for 100% of patients. The coders obtained perfect reliability for this theme (ICC = 1.0).

The second theme found in the narrative for Transition Visit 1 was that the CCLS conducted an educational assessment and identified if the patient understood their disease status. For example, CCLS documented, "Pt has a basic understanding of her disease as evidenced by knowing her PCP, no transfusion history, and no surgeries." In addition, CCLS also documented statements such as, "Pt has a good understanding of his disease as he knows his medications and type of sickle cell (hgb SS). Areas in which he needs encouragement include goals, school plans, and other life skills." Furthermore, in another patient note CCLS documented, "CCLS discussed
pt's medical history and completed form. Overall, pt needs some support with basic understanding of type of sickle cell, his medications, and baseline hgb and O2. He has a pediatrician and has his own phone. He knows his medications and how much HU [hydroxyeruea] he should take daily, but cannot elaborate on folic acid or aspirin. He could greatly benefit from medication education." The CCLS also identified if the patient knew their health insurance and asked if they had a driver’s license or access to a vehicle. In another patient’s EMR, a CCLS documented, "Pt knows little about his health history apart from his allergies, medications, and surgeries. He does not know his sickle cell type, PCP, complete understanding and dosage of medications, or baseline hemoglobin or oxygen. He also does not have a car, driver’s license, or know his health insurance. His lack of knowledge was validated and he was told not to get discouraged as this is why the Transition Clinic is in place." The CCLS documented an educational assessment in 38% of chart notes. The coders obtained perfect reliability for this theme (ICC = 1.0).

The third theme found in the narrative for Transition Visit 1 was that the CCLS documented provision of an "All About Me" form for 77% of patients. Appendix F. shows an example of the "All About Me" form used. The "All About Me" form included the following: "What is your favorite thing to do in your free time?" and "What words best describe you?" The CCLS documented the completion of the "All About Me" form in 77% of chart notes. The coders obtained perfect reliability for this theme (ICC = 1.0).

The fourth theme found in the narrative for Transition Visit 1 was rapport building among the CCLS and the patient. CCLSSs use rapport building to develop a trusting a relationship with the patient and their family. Examples of how CCLSSs use rapport building include the following: inquiring about the patient’s interests, school, future plans, and family life. The CCLS
inquired who the patient lives with, what grade they are in and if they have future plans (e.g., work, attend college). For example, "Pt describes herself as goofy, outgoing, and stylish. She is "gawdy" and likes makeup, nails, etc." was documented in the EMR. In another patient’s EMR, CCLS documented, "They have plans to attend college, probably somewhere close. Pt does not work. He likes to play on his PS4. He describes himself as a mama's boy, a clown, and funny." The CCLS documented rapport building in 85% of chart notes. The coders obtained moderate reliability for this theme (ICC = 0.625). A summary of the typical Transition Visit 1 details may be found in Table 1.
Table 1. Transition Visit 1 - Certified Child Life Specialist interventions, prevalence among chart notes, definitions, examples, and reliability

<table>
<thead>
<tr>
<th>CCLS Interventions</th>
<th>Prevalence</th>
<th>Definition</th>
<th>Example</th>
<th>ICC</th>
</tr>
</thead>
<tbody>
<tr>
<td>Introduction of self and services</td>
<td>100%</td>
<td>CCLS introduced themselves and provided the patient with an introduction of child life services, which included the role of a CCLS, how the CCLS can best support the patient in transition process, and education related to the program and child life role.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Educational Assessment</td>
<td>38%</td>
<td>CCLS identified if the patient understood their disease status.</td>
<td>&quot;CCLS discussed pt’s medical history and completed form. Overall, pt needs some support with basic understanding of type of sickle cell, his medications, and baseline hgb and O2. He has a pediatrician and has his own phone. He knows his medications and how much HU [hydroxyurea] he should take daily, but cannot elaborate on folic acid or aspirin. He could greatly benefit from medication education.&quot;</td>
<td></td>
</tr>
<tr>
<td>All About Me</td>
<td>92%</td>
<td>CCLS documented provision of &quot;All About Me&quot; form.</td>
<td>Questions include: &quot;What is your favorite thing to do in your free time?&quot; and &quot;What words best describe you?&quot;</td>
<td>1.0</td>
</tr>
<tr>
<td>Rapport Building</td>
<td>85%</td>
<td>CCLS use rapport building to develop a trusting relationship with the patient and their families.</td>
<td>&quot;Pt describes herself as goofy, outgoing, and stylish. She is &quot;gawdy&quot; and likes makeup, nails, etc.&quot;</td>
<td>0.625</td>
</tr>
</tbody>
</table>

*n = 13 for child life transition visit chart notes. Intraclass correlation coefficient (ICC) was used to measure reliability among coder*
Child Life Intervention During SCD Transition Visit 2

To answer the research question at Transition Visit 2, 18 EMRs were accessed and only 13 chart notes ($n = 13$) were found to have CCLS chart notes for Transition Visit 2. There were five missing chart notes. Results of the content coding and thematic analysis by the two coders indicated four categories of CCLS interventions during Transition Visit 2 for patients with SCD: (1) patient education, (2) family planning, (3) provision of handouts, and (4) educational assessment.

Patient education was a primary theme evident in the narrative for Transition Visit 2 in the sickle cell transition clinic. The CCLS reviewed genetic inheritance of SCD, including a brief explanation of Punnett squares (i.e., a square diagram used to display genetic outcomes using phenotype combinations) and education related to genetics. The CCLS introduced an activity consisting of sickle cell dice and assessed whether the patient was able to complete the Punnett square with no, minimal, or moderate help. For example, CCLS documented, "Brief explanation of Punnett square and genetics were presented to pt...pt instructed that outcomes of the square must be observed for each pregnancy." The CCLS documented patient education in 100% of chart notes. The coders obtained perfect reliability for this theme (ICC = 1.0).

The second theme found in the narrative of Transition Visit 2 was family planning. During the educational intervention, the CCLS documented that explanation was given to the patient that the outcomes of the Punnett square must be observed for each pregnancy. For example, CCLS documented, "Pt’s mother stated that he currently has a girlfriend whose family has a history of sickle cell disease. The girlfriend has sickle cell trait, per mom. Pt’s mother asked CCLS to emphasize that there is a possibility he could have a child with sickle cell disease should he and his current girlfriend continue their relationship further." The family planning
theme was present in 85% of the chart notes. The coders obtained perfect reliability for this theme (ICC = 1.0).

The third theme found in the narrative of Transition Visit 2 was the documented provision of educational handouts, such as Know Your Sickle Cell Status (K.Y.S.S.), Genes for Teens, Types of SCD, and Sickle Cell Disease and Trait, in 100% of the patient chart notes. K.Y.S.S. is a pamphlet used to provide education on SCD and SCT (Appendix G). The pamphlet includes a glossary of SCD related terms, educational activities and puzzles, a Punnett square depicting genetic inheritance, common types of sickle cell, and symptoms of SCT. Genes for Teens with SCD also provides information related to genetic inheritance (Appendix H). Types of SCD is an education resource that explains types of SCD, including differences in disease status and SCD related facts (Appendix I). Sickle Cell Disease and Trait defines sickle cell anemia and addresses misconceptions associated with SCD (Appendix J). In addition, it provides information about diagnosis and newborn screening. The coders obtained perfect reliability for this theme (ICC = 1.0).

The fourth theme found in the narrative for Transition Visit 2 was an educational assessment, and this category was present in 100% of the reviewed charts. Review of the child life documentation in the EMR demonstrated that the CCLS conducted an educational assessment during Transition Visit 2, reviewing sickle cell status, genetic inheritance, and types of SCD and SCT while providing patient specific education. For example, CCLS documented,"With minimal help, pt rolled SCD dice and completed a few Punnett squares with CCLS." The coders obtained perfect reliability for this theme (ICC = 1.0). A summary of the typical Transition Visit 2 details may be found in Table 2.
Table 2. Transition Visit 2 - Certified Child Life Specialist interventions, prevalence among, definitions, examples, and reliability

<table>
<thead>
<tr>
<th>CCLS Interventions</th>
<th>Prevalence</th>
<th>Definition</th>
<th>Example</th>
<th>ICC</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient Education</td>
<td>100%</td>
<td>The CCLS reviewed genetic inheritance of SCD, including a brief explanation of Punnett squares and education related to genetics. The CCLS introduced an activity consisting of sickle cell dice.</td>
<td>&quot;Brief explanation of Punnett square and genetics were presented to pt…Pt instructed that outcomes of the square must be observed for each pregnancy.&quot;</td>
<td>1.0</td>
</tr>
<tr>
<td>Family Planning</td>
<td>85%</td>
<td>Explanation was given to the patient that the outcomes of the Punnett square must be observed for each pregnancy</td>
<td>&quot;Pt’s mother stated that he currently has a girlfriend whose family has a history of sickle cell disease. The girlfriend has sickle cell trait, per mom. Pt’s mother asked CCLS to emphasize that there is a possibility he could have a child with sickle cell disease should he and his current girlfriend continue their relationship further.&quot;</td>
<td>1.0</td>
</tr>
<tr>
<td>Provision of handouts</td>
<td>100%</td>
<td>CCLS documented provision of handouts.</td>
<td>K.Y.S.S., Genes for Teens, Types of SCD, and Sickle Cell Disease and Trait</td>
<td>1.0</td>
</tr>
<tr>
<td>Educational Assessment</td>
<td>100%</td>
<td>CCLS assessed whether the patient was able to complete the Punnett square with no, minimal, or moderate help.</td>
<td>&quot;With minimal help, pt rolled SCD dice and completed a few Punnett squares with CCLS.&quot;</td>
<td>1.0</td>
</tr>
</tbody>
</table>

\(n = 13\) for child life transition visit chart notes. Intraclass correlation coefficient (ICC) was used to measure reliability among coders.
**Child Life Intervention During SCD Transition Visit 3**

To answer the research question at time 3, 18 EMRs were accessed and 11 chart notes \((n=11)\) were found to have CCLS chart notes for Transition Visit 3. There were seven missing chart notes. Results of the content coding and thematic analysis by the two coders indicated four categories of CCLS interventions during Transition Visit 3 for patients with SCD: (1) discussion and review of pain plan, (2) education related to non-pharmacological pain management, (3) deep breathing demonstration, and (4) guided imagery demonstration.

Discussion and review of pain plan was the first theme present in the narrative for Transition Visit 3. The CCLS documented inquiry about the patient’s pain plan in 82% of the reviewed charts. Discussion and review of the pain plan was documented, including methods of pain management. For example, CCLS documented, "Pt described their pain plan as taking a bath, drinking fluids, rest and taking medications." In another chart note, CCLS documented, "Patient described their pain plan as taking a bath and taking medications (ibuprofen)." CCLS documented in another patient’s EMR, "Pt described his pain plan as taking medications and using a heating pad." The coders obtained perfect reliability for this theme \((ICC = 1.0)\).

Inquiring about the patient’s pain plan allowed the CCLS to then introduce non-pharmacological pain management. The second theme present in the narrative of Transition Visit 3 was patient education related to non-pharmacological pain management. Education related to non-pharmacological pain management was a theme present in 100% of the chart notes. For example, the CCLS documented introduction of "deep breathing and guided imagery to be used in conjunction with her pain plan." The coders obtained perfect reliability for this theme \((ICC = 1.0)\).
The third theme present in the narrative of Transition Visit 3 was a deep breathing demonstration. The CCLS documented that a deep breathing demonstration was an intervention during Transition Visit 3, and documented patient engagement in the demonstration. For example, CCLS documented, "After introducing deep breathing, pt took deep breaths, but remained silent." The CCLS documented deep breathing demonstration in 91% of the reviewed chart notes. The coders obtained perfect reliability for this theme (ICC = 1.0).

The fourth theme found in the narrative for Transition Visit 3 was a guided imagery demonstration. The CCLS documented that a guided imagery demonstration was an intervention during Transition Visit 3. For example, CCLS documented, "Pt did participate in guided imagery intervention. After completing guided imagery session, pt stated he was "very relaxed." CCLS commended pt on his participation and encouraged him to use it during times of stress, anxiety, or pain." In another patient’s Transition Visit 3 note, CCLS documented, "When asked about a happy place for guided imagery, he shrugged his shoulders and with time suggested the beach. Pt minimally participated in guided imagery intervention." A guided imagery demonstration was documented in 91% chart notes. The coders obtained perfect reliability for this theme (ICC = 1.0). A summary of the typical Transition Visit 3 details may be found in Table 3.
Table 3. Transition Visit 3 - Certified Child Life Specialist interventions, prevalence among, definitions, examples, and reliability

<table>
<thead>
<tr>
<th>CCLS Interventions</th>
<th>Prevalence</th>
<th>Definition</th>
<th>Example</th>
<th>ICC</th>
</tr>
</thead>
<tbody>
<tr>
<td>Discussion and review of pain plan</td>
<td>82%</td>
<td>Discussion and review of the pain plan was documented, including methods of pain management.</td>
<td>&quot;Pt described their pain plan as taking a bath, drinking fluids, rest and taking medications.&quot;</td>
<td>1.0</td>
</tr>
<tr>
<td>Education related to nonpharmacological pain management</td>
<td>100%</td>
<td>CCLS introduced nonpharmacological pain management to pt.</td>
<td>CCLS documented introduction of &quot;deep breathing and guided imagery to be used in conjunction with her pain plan.&quot;</td>
<td>1.0</td>
</tr>
<tr>
<td>Deep breathing demonstration</td>
<td>91%</td>
<td>CCLS documented engagement in demonstration.</td>
<td>&quot;After introducing deep breathing, pt took deep breaths, but remained silent.&quot;</td>
<td>1.0</td>
</tr>
<tr>
<td>Guided imagery demonstration</td>
<td>91%</td>
<td>CCLS documented engagement in demonstration.</td>
<td>CCLS documented, &quot;Pt did participate in guided imagery intervention. After completing guided imagery session, pt stated he was &quot;very relaxed.&quot; CCLS commended pt on his participation and encouraged him to use it during times of stress, anxiety, or pain.&quot;</td>
<td>1.0</td>
</tr>
</tbody>
</table>

\( n = 11 \) for child life transition visit chart notes. Intra-class correlation coefficient (ICC) was used to measure reliability among code
DISCUSSION

The purpose of this retrospective chart review was to analyze medical notes pertaining to SCD and child life services and identify the child life interventions provided for AYA patients during the first three transition visits with the healthcare team in a SCD transition clinic by the CCLS. The findings from the retrospective chart review were not surprising and the interpreted themes were guided by the literature and expertise of the CCLSs interpreting the data. The findings from the qualitative thematic analysis revealed that the CCLS encouraged the AYA patients to be active participants in their health care experience and provided the AYA patients with developmentally appropriate interventions to assist in meeting their psychosocial needs during the transition process.

Findings from this study identified child life interventions during the three transition visits for patients with SCD. Findings suggested that CCLSs provide an introduction of self and services and spend time during the initial transition building rapport and making an educational assessment. Patients with SCD are followed by the health care team to provide appropriate treatment and maintenance of disease-specific symptoms. Therefore, it is possible that patients with SCD have had an encounter with a CCLS during a health care experience. The scope of child life practice can be broad and CCLSs have unique roles based on the needs of the patients and families. By introducing child life services in transition, CCLSs can clarify their role in the transition process. For example, the findings from this study show that the CCLS explained the role of child life practice during transition visits as support and education. The current child life literature supports the findings from this study related to diagnosis teaching and education. CCLSs receive education and training to make educational assessments and spend time getting to know the patient and building a trusting relationship (i.e., building rapport), which is important.
for patients with SCD (Boles et al., 2020; Clayton-Jones et al., 2021). Through building rapport, CCLSs can further advocate for patient- and family- centered care (Bell et al., 2009). The inter-rater reliability for the theme rapport building was not as robust as the other coefficients. This could have been confounded with the "All About Me" form and the coding decisions of the raters.

During Transition Visit 1, findings revealed that CCLSs implement resources and tools to assist in their interventions such as the "All About Me" form. The "All About Me" form asked questions related to the patient’s transportation. For sickle cell patients in the transitional program, transportation to appointments is an important component of a successful transition, and understanding risk and resilience factors is necessary when making a child life assessment (Beickert & Mora, 2017; Brennan-Cook et al., 2018). The "All About Me" form included who the patient lives with, what grade they are in and if they have future plans (e.g., work, attend college, trade school). The information outlined on the "All About Me" form further contributes to the focus of care (Goldberger et al., 2009). Findings revealed that the "All About Me" form was used during Transition Visit 1 to allow the CCLS the opportunity to learn about the patient and patient’s family while building rapport with the patient. The "All About Me" form identified potential barriers that the patient may experience when planning for transportation to and coordination of doctor’s appointments (e.g., inquiring about the patient’s driver’s license and caregiver presence and support).

Current literature pertaining to CCLSs’ assessments support the findings from the retrospective chart. Koller (2008) found that CCLSs’ assessments include child variables, family variables, illness variables, and medical experiences. CCLSs must consider variables that contribute to AYA patient’s coping and adjustment during the transitional process from pediatric
care to adult management. Cultural considerations and social factors are included in a CCLS’s psychosocial assessment (Boles et al., 2020; Hollen & Skinner, 2009). Findings revealed that characteristics involving the individual’s immediate environment (i.e., caregiver presence, school, job) were documented in the patient’s EMR, and that CCLS’s assessments include how AYA patients with SCD were shaped by external levels of influence, as would be expected based on Bronfenbrenner’s (1977) ecological theory.

Fenton et al. (2015) described family cohesion as an important predictor of a patient’s transition outcome. By learning about the patient and the patient’s family, the CCLS can identify potential risk and resilience factors associated with SCD, such as fear, lack of communication, and mistrust of health care providers (Long et al., 2011; Sobota et al., 2014). The findings from this study support the importance of CCLSs and family centered care when working with AYA patients with SCD.

Findings from the retrospective chart review of child life notes during Transition Visit 2 reveal educational assessment as a theme for Transition Visit 2. The findings suggested that an educational assessment is ongoing and that CCLSs use their assessments to plan and implement patient specific interventions to support the focus of care (Goldberger et al., 2009). Hollen and Skinner’s (2009) literature pertaining to how CCLSs make assessments supports the findings from this study that child life assessments are ongoing and may change over time. For example, patient education was a common theme during Transition Visit 2. CCLS provided education related to genetic inheritance. Introduction of education allowed the CCLS the opportunity to make an educational assessment and inform more about family planning. CCLSs provide information related to family planning, including future goals and aspirations. Understanding a patients’ wishes is an important component of PFCC (Bell et al., 2009). In addition, family
planning education is an important component of education related to diagnosis and screening of SCD (Stevens et al., 2019). The lack of follow-up care and communication related to diagnosis and screening of SCD is concerning (Stevens et al., 2019). Discussion and education provided by a CCLS provides the patient the opportunity to be informed about SCD and SCT before transitioning to adult management.

Children and adolescents with chronic illness can experience hopelessness and despair associated with their illness that can impact planning for the future (Boles et al., 2020; Goralski et al., 2017). CCLSs can assist with goal setting and help to create action plans for the patient to support the focus of care (Goldberger et al., 2009). In this study, the CCLS also documented provision of disease specific resources and handouts during Transition Visit 2. Findings suggested that CCLSs are equipped with knowledge and understanding of developmentally appropriate and disease specific resources to aid in their patient interventions, as found by Boles et al. (2020). The findings from the study identified that CCLSs provide educational interventions to address disease-specific needs for AYA patients with SCD.

Several other categories were present during the retrospective chart review of child life notes for Transition Visit 3. The CCLS initiated discussion and inquired about the patient’s pain plan. Treatment and maintenance of pain is important for patients with SCD (CDC, 2020). Barakat et al. (2006) found that daily management and treatment for SCD can interfere with daily functioning. Identification of a patient’s pain plan allows the CCLS and patient to form a coping plan that includes considerations for school, daily activities, and patient specific interests. Psychological stress can enhance physiological symptoms, therefore CCLS facilitating discussion of a patient’s daily life and expectations is helpful in understanding a patient’s pain plan (Shah et al., 2020).
Also during Transition Visit 3, the CCLS introduced non-pharmacological pain management and provided education related pain management. In addition, the CCLS documented that deep breathing and guided imagery demonstrations were introduced. Child life literature stated that CCLSs provide preparation, support, and model coping plans for patients (Koller, 2008; Sisk, 2016; Thompson, 2009). By understanding a patient’s pain plan, CCLSs can identify any misconceptions that need to be addressed by the health care team associated with treatment and maintenance of SCD related symptoms and pain. CCLSs can assess a patient’s coping to develop a coping plan for future pain episodes and provide the patient with nonpharmacological methods that assist in meeting the goals and expectations for the patient to manage disease-related symptoms and optimize daily functioning.

CCLSs should consider the findings from this retrospective chart review as possible interventions to implement with patients with SCD. The findings from this study were not surprising based on the current literature related to SCD and child life practices. Current literature related to child life implementation of disease-specific interventions supports the findings from this study. The disease-specific interventions found in the qualitative data analysis align with common child life practices. Child life interventions are commonly cited throughout literature related to the child life profession, scope of practice, and patient and family centered care, but this study identified disease-specific interventions that align with the CCLS’s assessments of AYA patients with SCD. The interventions implemented for patients with SCD included patient specific assessments, but the interventions themselves were not patient specific. Instead findings revealed that interventions were disease-specific (i.e., education related to SCD, genetic inheritance, nonpharmacological pain management) and included patient specific psychosocial assessments. The findings from this study identified tools and resources (e.g.,
educational handouts) to be updated and reviewed periodically to ensure that the CCLS is providing the patient and family with the most up to date disease-specific information available.

**Implications for Practice**

The findings from this study provide implications for child life practice, specifically for CCLTs who provide interventions for AYA patients with SCD in preparation for their transition to adult management. CCLTs can provide introduction of self and services, conduct an educational assessment, and facilitate rapport building during an initial visit with AYA patients with SCD to inform the patient and family of the role of child life practice during the transition process. Furthermore, implementation of an "All About Me" form can be used to identify patient and family needs, and facilitating rapport building can be used to enhance the relationship between the CCLS and the patient and family. CCLTs can also provide AYA patients with handouts and resources to address disease specific topics. Furthermore, CCLTs may consider discussion and review of a patient’s pain plan, and provide education and demonstrations related to nonpharmacological pain management for AYA patients with SCD (e.g., deep breathing and guided imagery) to assist in development of a coping plan for each AYA patient with SCD.

In addition, CCLTs can provide the patient with education and information related to family planning as part of their interventions. CCLTs can continue their educational assessment during each patient encounter to assist in a patient’s understanding of their disease status and treatment. Documentation of an educational assessment was only present in 38% of notes compared to introduction of self and services (100%), "All About Me" form (92%), and rapport building (85%) for Transition Visit 1. Child life documentation of interventions, including an educational assessment, could be used to identify a patient’s understanding of their disease status, contribute to future educational opportunities, and improve patient outcomes. Child life
documentation and communication is an important component of child life services and continuity of care for patients and families.

The results from this study are informative for CCLTs and other health care professionals. The results detail information about child life interventions and can be used as a framework to inform the role of a CCLS. Awareness of the child life role, child life interventions, and child life responsibilities can provide an opportunity for collaboration among the multidisciplinary team, further advance patient care outcomes (e.g., trust of health care providers, rapport building with health care team), and enhance a patient’s experience in their health care environment through communication and collaboration. CCLTs can also benefit from additional disease-specific education for their unit of coverage in addition to their education and training.

Furthermore, application and understanding of Bronfenbrenner’s ecological framework can assist CCLTs in their interventions with AYA patients and families because it identifies levels of external influence and provides perspective on an AYA patient’s interpersonal relationships with others and their environment. CCLTs can use Bronfenbrenner’s (1977) ecological theory to help shape their patient and family assessments. Application of Bronfenbrenner's ecological framework can be used to understand the spheres of influence (e.g., home environment, resources, patient's goals for the future). CCLTs can conduct a thorough psychosocial assessment to ensure that each transition visit is individualized to the patient's specific needs and that child life interventions are adapted based on each patient and family.

Furthermore, regarding nonpharmacological pain management, findings from the study include discussion of a patient's pain plan and what the pain plan includes (e.g., opportunity for rest and purchasing essential oils and diffusers). The CCLS must consider how realistic it is for the
patients to follow suggestions such as using alternative methods of pain management. CCLSs can also expand on disease-specific education related to SCD and SCT to assist in future family planning goals that can differ among patients.

Due to the design of this retrospective chart review, documentation was an essential component of this study. Findings from this study indicated that CCLSs provide documentation in the EMR detailing their patient interventions. Documentation is an important responsibility of child life professionals (Hollen & Skinner, 2009). However, there was missing data for each transition visit. Perhaps this could be due to lack of child life presence, or CCLSs may not document every patient intervention. The reason is unclear, and future research is needed to address the questions related to child life documentation requirements, accountability, frequency of documentation, and EMR accessibility for CCLSs.

There is also an opportunity for future research focusing on the patient experience and patient narrative that could be used to build a per patient case narrative across time and identify how CCLSs build upon each patient visit. Therefore, focusing on the patient’s experience over time in the transition clinic could enhance understanding of child life interventions and provide further implications for patient care and child life interventions.

**Limitations**

There were several limitations to consider when interpreting the results of this study. The small sample size limited the generalizability of the results. In addition, the sample was collected from one outpatient clinic in the southeastern United States. The retrospective chart review was used to analyze data from a CCLS in one outpatient clinic, therefore practices could be limited based on the model that was established for this transition clinic. Results could vary based on a CCLS’s practices and among other outpatient hematology and oncology clinics. Child life
practice could differ based on the setting in which the patients are seen (e.g., inpatient or outpatient) and resources that are available for the CCLS (i.e., if there is a designated multidisciplinary team for patients with SCD). Results could differ based on the timing in which the data from the retrospective chart review was collected. A larger sample would have provided more detailed results, including greater insight on child life interventions. There was also missing data from the retrospective chart review, which limited the number of charts that could be reviewed for each transition visit. It is possible that not all child life interventions were outlined in the EMR. Perhaps, if all child life interventions were not documented in detail for each visit, the retrospective chart review did not fully capture the scope of child life practice for AYA patients with SCD. The missing data did not affect the outcome of the study. The educational assessment theme during Transition Visit 1, which had a lower prevalence, might have been more prevalent if more data was available for Transition Visit 1. Completing the retrospective chart review for a series of more transition visits over a longer period of time could increase findings and provide better detail as to what CCLSs do for AYA patients with SCD.
CONCLUSION

The findings from this retrospective chart review identified that CCLSS provided interventions for AYA patients with SCD during Transition Visit 1, Transition Visit 2, and Transition Visit 3. The purpose of this retrospective chart review was to analyze medical notes pertaining to SCD and child life services. The qualitative thematic analysis identified the following child life interventions for AYA patients with SCD: introduction of self and services, educational assessments, an "All About Me" form, rapport building, patient education, family planning, provision of handouts, discussion and review of pain plan, education related to nonpharmacological pain management, deep breathing demonstration, and guided imagery demonstration. Current literature related to child life practice supports the findings from this study. The findings from this study can be used to further identify and support the role of CCLSS working with AYA patients with SCD. Results demonstrate that child life interventions vary based on the transition visit type. However, due to the nature of this retrospective chart review, the importance of child life documentation was discovered. Implications from this study can be used to advance the delivery of child life interventions and address the goals of care for AYA patients with SCD in the transitional process from pediatric care to adult management.
Appendix A.

IRB Approval Letter

Institutional Review Board (IRB)
IORG#: IORG0004976

Date: August 18, 2021

Study Number: 2021-115
Study Title: What do Child Life Specialists do for Sickle Cell Patients Transitioning from Pediatric Health Care to Adult Management?
Primary Investigator: Amanda Stark, B.S.
Secondary Investigator(s): Brittany Wittenberg, Ph.D., M.S., B.S., Jessica Templet
Primary Reviewer: Leslie S Son, PhD
SecondaryReviewer: Katie Vance, PhD
Exemption Category: (4) Secondary research for which consent is not required.

Dear Ms. Stark,

Please accept this as documentation of review and approval of the above referenced research proposal by the Franciscan Missionaries of Our Lady University Institutional Review Board. This study qualifies as Exempt under the following Revised Common Rule category: (4) Secondary research for which consent is not required.

This study does not have an expiration date. However, if you plan to make any significant changes in the conduct of this research, you are required to submit a Request for Amendment form and receive IRB approval before you proceed with the changes.

Please notify the IRB when your research is complete or cancelled by submitting a Closure Notification form.

Thank you for your submission and I would like to wish you success with your project.

Best regards,

Dr. Michael T. Dreznick, Associate Professor
Franciscan Missionaries of Our Lady University and IRB Chair
Appendix B.

IRB Authorization Agreement


Name of Institution or Organization Providing IRB Review (Institution/Organization A):
Franciscan Missionaries of Our Lady University

IRB Registration #: IORG0004976 Federalwide Assurance (FWA) #, if any: FWA00011379

Name of Institution Relying on the Designated IRB (Institution B):
Louisiana State University

FWA #: FWA00003892

The Officials signing below agree that Louisiana State University may rely on the designated IRB for review and continuing oversight of its human subjects research described below. (check one)

(____) This agreement applies to all human subjects research covered by Institution B’s FWA.

(____) This agreement is limited to the following specific protocol(s):

Name of Research Project: What do Child Life Specialists do for Sickle Cell Patients Transitioning from Pediatric Health Care to Adult Management?
Name of Principal Investigator: Amanda Stark
Sponsor or Funding Agency: Award Number, if any:

(____) Other (describe):

The review performed by the designated IRB will meet the human subject protection requirements of Institution B’s OHRP-approved FWA. The IRB at Institution/Organization A will follow written procedures for reporting its findings and actions to appropriate officials at Institution B. Relevant minutes of IRB meetings will be made available to Institution B upon request. Institution B remains responsible for ensuring compliance with the IRB’s determinations and with the Terms of its OHRP-approved FWA. This document must be kept on file by both parties and provided to OHRP upon request.

Signature of Signatory Official (Institution/Organization A):

Print Full Name: Bro. Edward A Violett Institutional Title: Provost & Vice President for Academic Affairs

NOTE: The IRB of Institution A may need to be designated on the OHRP-approved FWA for Institution B.

Signature of Signatory Official (Institution B):

Print Full Name: Stephen Beck Institutional Title: Associate Vice President

Date: 12/13/21
Appendix C.
Child Life Outpatient Hematology/Oncology Transition Visit 1

Visit Type:

Goal(s):

Psychosocial Assessment

Communications:

Behaviors:

Interventions

Patient intervention:

Family interventions:

CCLS met with __________ and {Family Member:_________} to introduce self and services during Transition Clinic Visit 1.

All About Me form completed. Pt {DOES/DOES NOT} know their health insurance. Pt {DOES/DOES NOT} have their driver's license. Pt {DOES/DOES NOT} have access to a vehicle. Pt lives in *** with {________}. __________ goes to ***school and is in the 1st-12th grade. They {:"have plans to attend","do not have plans to attend"} {"college","trade school"}. Pt {DOES/DOES NOT} work.

Pt favorites include:

- 
- 
-
Pt/family denied any questions or concerns at this time. CCLS will remain available to patient and family as needed or as warranted by changes in patient and family status.

Plan:

Child Life Services Plan:

Outcome/Follow-up:

Patient Education:

Time spent:

CCLS

@TODAYDATE@
Appendix D.

Child Life Outpatient Hematology/Oncology Transition Visit 2

Visit Type:

Goal(s):

Psychosocial Assessment

Communications:

Behaviors:

Interventions

Patient intervention:

Family interventions:

CCLS followed up with _________ and {Family Member} during Transition Visit 2 to review genetics of sickle cell disease. Brief explanation of Punnett square and genetics were presented to patient. With {no/min/mod} help,__________ rolled sickle cell dice and completed a Punnett squares with CCLS. Patient instructed that outcomes of the square must be observed for each pregnancy. Patient { does/does not} want to have children.

__________ provided with the following handouts: K.Y.S.S., Genes for Teens, Types of SCD, and Sickle Cell Trait.
Patient/family denied any questions or concerns at the conclusion of the visit. CCLS will remain available to patient and family as needed or as warranted by changes in patient and family status.

Plan:

Child Life Services Plan:

Outcome/Follow-up:

Patient Education:

Time spent:

CCLS

@TODAYDATE@
Appendix E.
Child Life Outpatient Hematology/Oncology Transition Visit 3

Visit Type:

Goal(s):

Psychosocial Assessment

Communications:

Interventions

Patient intervention:

Family interventions:

CCLS followed up with __________ and {Family Member} during Transition Visit 3 to review non-pharmacologic methods of pain management. Patient described their pain plan as {"taking a bath","drinking fluids","rest","taking medications"}. CCLS introduced deep breathing and guided imagery to be used in conjunction with patient's pain plan. Patient {did/not} engage in deep breathing demonstration. Patient {did/not} participate in guided imagery intervention.

Patient/family denied any questions or concerns at the conclusion of the visit. CCLS will remain available to patient and family as needed or as warranted by changes in patient and family status.

Plan:

Child Life Services Plan:

Outcome/Follow-up:
Patient Education:

Time spent:

CCLS

@TODAYDATE@
Appendix F.

All About Me
Transition Clinic

Full name: ____________________ Nickname: ________________
Birthday: ________________ Where were you born: ________________
What city do you live in?

Who lives in your house? Any brothers or sisters (names, birthdates)?

Any pets? (Kinds, names)

Favorite:
  Food-
  Candy-
  Cake or cookie cake?

What words best describe you?

Where do you go to school? What grade are you in? Favorite class?

What are your plans for after school? College? Trade school? Straight to a job?

What do you want to be when you grow up?

How would you feel about living by yourself?

Do you have a driver’s license? What are your plans for transportation?

What type of health insurance do you have? What is the name of your plan?

Favorite thing to do in your free time?

Date Completed: ______________
Appendix G.

Know Your Sickle Status!
And spread the knowledge!
Do you KYSS (Know Your Sickle Status)?

Does anyone in your family have Sickle Cell Trait or Sickle Cell Disease?
Who can get Sickle Cell Disease?

Sickle Cell Disease (SCD) affects people of all different racial and ethnic backgrounds.

For this reason, in most U.S. hospitals, all babies are now screened at birth for Sickle Cell Disease.
What is the origin of Sickle Cell Disease?

Sickle Cell has origins in:
... tropical areas of the world where *malaria* was common.

People with Sickle Cell Trait have an increased protection against malaria, which is spread by mosquitoes.
What is the ratio of Sickle Cell Trait?

1 in 12 African Americans have Sickle Cell Trait.

1 in 100 Latinos have Sickle Cell Trait.
What is Sickle Cell Trait?

People who have Sickle Cell Trait inherit:

Normal Gene  Sickle Cell Gene
Sickle Cell Trait is a genetic “carrier” condition.

Sickle Cell Trait is only diagnosed with special blood tests such as Hemoglobin Electrophoresis.
Are there health problems associated with Sickle Cell Trait?

People with Sickle Cell Trait usually Do NOT have health problems caused by the trait.

HOWEVER

People with Sickle Cell Trait may have problems under certain conditions such as:

- Exhaustion
- Dehydration
- Hypoxia (low oxygen)
What is Sickle Cell Disease?

Sickle Cell Disease (SCD) is a group of blood disorders that can be very serious. It is a genetic disorder. BOTH parents must pass on an abnormal gene for a child to be born with the disease.
Hemoglobin SS is the most common type of Sickle Cell Disease in the U.S. Both parents must have Sickle Cell Trait for their child to be born with Hemoglobin SS.

Other Common Forms of Sickle Cell Disease include:

- Hemoglobin SC
- Sickle Beta 0 Thalassemia
- Sickle Beta + Thalassemia
Sickle Cell Disease affects the *hemoglobin* part of the blood. Hemoglobin carries oxygen throughout the body. Normal red blood cells are round, soft, smooth and move easily throughout the body.
Sickle red blood cells become hard, sticky and look like a sickle or a banana.

When the sickle cells travel through blood vessels, they can get stuck and clump together. This can cause pain and other problems.
What are some symptoms of Sickle Cell Disease?

Pain
Anemia
Increased Infections
Leg Ulcers
Strokes
Organ Damage
Delayed Growth
Yellow Jaundice
How do you inherit Sickle Cell Disease?

If both parents have sickle cell trait there is a 25% chance that their child will have sickle cell disease.
With EACH pregnancy, the chances for Sickle Cell Trait and Sickle Cell Disease remain the same.
Why is testing important?

It is important to identify people with Sickle Cell Trait, so they will be aware of their risk of having children with Sickle Cell Disease.

Get tested and...

Know Your Sickle Status!

K.Y.S.S.
<table>
<thead>
<tr>
<th>Term</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>carrier</td>
<td>an individual who carries a gene</td>
</tr>
<tr>
<td>dehydration</td>
<td>lack of water in the body</td>
</tr>
<tr>
<td>exhaustion</td>
<td>very tired</td>
</tr>
<tr>
<td>genetic</td>
<td>inherited from parents</td>
</tr>
<tr>
<td>hemoglobin</td>
<td>gives blood its red color and carries oxygen from the lungs to the rest of the body</td>
</tr>
<tr>
<td>hemoglobin electrophoresis</td>
<td>a blood test that can diagnose sickle cell disease and trait</td>
</tr>
<tr>
<td>hypoxia</td>
<td>low oxygen</td>
</tr>
<tr>
<td>jaundice</td>
<td>yellowing of the skin and eyes</td>
</tr>
<tr>
<td>malaria</td>
<td>a potentially fatal infectious disease spread by mosquitoes</td>
</tr>
<tr>
<td>ulcers</td>
<td>sores</td>
</tr>
</tbody>
</table>
## KYSS Kards

Match the questions with the correct answers.

<table>
<thead>
<tr>
<th>Questions</th>
<th>Answers</th>
</tr>
</thead>
<tbody>
<tr>
<td>1) Who can get sickle cell disease?</td>
<td>A. 1 in 12 African Americans have sickle cell trait</td>
</tr>
<tr>
<td>2) What is the approximate ratio of African Americans that are sickle cell trait carriers?</td>
<td>B. Yes</td>
</tr>
<tr>
<td>3) If you have Sickle Cell Trait, you have one SickleCell Gene and one ______ Gene.</td>
<td>C. Sickle Cell Disease (SCD) affects people of all different racial and ethnic backgrounds</td>
</tr>
<tr>
<td>4) If both parents have sickle cell Trait, they can have a child with Sickle Cell Disease.</td>
<td>D. 50 % (or 2 in 4 chance)</td>
</tr>
<tr>
<td>5) If both parents have sickle cell trait, what is the percentage of chance that the child will have trait?</td>
<td>E. 25 % (or 1 in 4 chance)</td>
</tr>
<tr>
<td>6) If both parents have sickle cell trait, what is the percentage of chance that the child will have sickle cell disease?</td>
<td>F. It is important to identify people with sickle cell trait, so they will be aware of their risk of having children with sickle cell disease.</td>
</tr>
<tr>
<td>7) If both parents have sickle cell trait, what is the percentage of chance that the child will have normal hemoglobin?</td>
<td>G. normal</td>
</tr>
<tr>
<td>8) Are these percentages the same with EACH pregnancy?</td>
<td>H. True</td>
</tr>
<tr>
<td>9) Why is sickle cell trait testing important?</td>
<td>I. 25 % (or 1 in 4 chance)</td>
</tr>
</tbody>
</table>
KYSS Square

What are the four possible outcomes for each child, if mom and dad both have Sickle Cell Trait?

What are the 4 possible outcomes for each child? Use the Punnett’s Square below to map them.

```
  | A | S |
---|---|---|
A |   |   |
DAD|   |   |
S  |   |   |
```
KYSS Krossword

Across

5  Point at which body has lost too much water
8  Different characteristics that can be passed on to offspring
9  Not usual or typical
10 To have or get from parents by the biological process of heredity
11 Yellowish in color; yellowing of the skin and the whites of the eyes caused by health problems
12 A chart used to show the possible gene combinations of two parents (2 words)
14 A sore in or on the body
16 Makes the blood red and carries oxygen in the blood
19 Passed from parents to offspring

Down

1 Extreme tiredness; weakening and loss of energy
2 What happens when red blood cells form a C-shape.
3 If both parents have sickle cell trait, the percentage of chance that the child will have trait
4 A person who “carries” a gene
6 Special blood test that can diagnose sickle cell trait or sickle cell disease (2 words)
7 If both parents have sickle cell trait, the percentage of chance that the child will have SCD (2 words)
13 When a person has a very low number of red blood cells in his/her body.
15 Abnormally low levels of oxygen in the cells
17 A potentially fatal, infectious disease passed along by the bite of an infected mosquito.
18 A trait that is inherited from parents
KYSS Krossword

Answers

SICK | HOSPITAL | EXHAUSTION

CAUTION | DEHYDRATION | BLOOD

ABNORMAL | INHERIT | Y FIVE

JAUNDICE | PUNNETTS SQUARE

ULCERS | P HYPOMEGALOGLLOBIN

GENESIS | ALANENIA
Interested in more information?

Contact:
St. Jude Children’s Research Hospital
Department of Hematology
by visiting
www.stjude.org/sickle cell
Appendix H.

If my partner has sickle cell disease, hemoglobin SS disease, will my children inherit sickle cell disease?

Yes, your children will inherit sickle cell disease. If you have sickle cell disease and have a child with a person who has sickle cell disease (hemoglobin SS disease), all of your children will have sickle cell disease.

![Gene Chart]

What will my children inherit?

- With each birth, what is your chance of having a child with sickle cell disease if your partner has sickle cell disease (SS)?
- With each birth, what is your chance of having a child with sickle cell disease if your partner has sickle cell trait (AS)?
- With each birth, what is your chance of having a child with sickle cell disease if your partner has normal hemoglobin (AA)?

Knowing your risk before you have a child will help you make wise decisions about family planning.
What are genes?
- Genes determine specific traits that are passed down to you or inherited from your parents. These traits are things like eye color, hair color, skin tone, physical features, and the type of hemoglobin in your blood.
- Genes are pieces of DNA that help make you who you are.
- You inherit one set of genes from your mother and one set of genes from your father.

What is sickle cell disease?
- Sickle cell disease affects part of the red blood cells, called hemoglobin.
- Hemoglobin helps carry oxygen from the lungs to the rest of the body.

How are genes related to sickle cell disease?
- Everyone has 2 hemoglobin genes. One gene is passed down from the mother and one is from the father.
- People with sickle cell disease inherit a hemoglobin S gene from one parent and another abnormal hemoglobin gene from the other parent.

What is sickle cell trait, and how is it different from the disease?
- People with sickle cell trait, called AS, inherit the normal hemoglobin A gene from one parent and an abnormal hemoglobin S gene from the other parent.
- Having the sickle cell trait does not mean you have sickle cell disease.
- If you have the trait, you can pass it on to your child.
- If you and your partner both have the trait, you might have a child with sickle cell disease.

Are there different types of sickle cell disease?
- Yes, sickle cell disease is a name for a group of disorders that have sickle hemoglobin. There are the most common types of sickle cell disease in the United States:
  - Hemoglobin SS disease
  - Sickle hemoglobin C disease (hemoglobin SC disease)
  - Two types of sickle beta thalassemia disease:
    - Sickle beta thalassemia disease (hemoglobin Sβ- disease)
    - Sickle beta zero thalassemia disease (hemoglobin Sβ0 disease)

How did I inherit sickle cell disease?
- People can inherit sickle cell disease in 3 different ways:
  - If both parents have sickle cell trait (AS) or
  - If one parent has sickle cell trait (AS) and the other parent has another abnormal hemoglobin gene.
  - If both parents have sickle cell disease.
- If you have hemoglobin SS disease, you inherited 2 hemoglobin S genes. one from each parent.
- If you have hemoglobin SC disease, you inherited a hemoglobin S gene from one parent and a hemoglobin C gene from the other parent.
- If you have hemoglobin Sβ disease, you inherited a hemoglobin S gene from one parent and a hemoglobin β gene from the other parent.

If my partner has sickle cell trait, will my children inherit sickle cell disease?
- Yes, your children may inherit sickle cell disease. If you have sickle cell disease, and your partner has sickle cell trait, with each birth there is a 1 out of 2 chance that your children will inherit a hemoglobin disease (50 percent chance). There is also a 1 out of 2 chance that your children will inherit a hemoglobin trait.

If my partner has normal hemoglobin A, will my children inherit sickle cell disease?
- No, if you have sickle cell disease and your partner has normal hemoglobin A, each child will inherit a hemoglobin trait. You will not have a child with sickle cell disease if your partner has normal hemoglobin A.
Appendix I.

Sickle Cell Patient Education Handout
St. Jude Baton Rouge Affiliate Clinic Educational Series

Genes for Teens
Types of Sickle Cell Disease
Teen/Young Adult
WHAT ARE THE DIFFERENT TYPES OF SICKLE CELL DISEASE?

There are many different types of sickle cell disease. The most common are:
- Hemoglobin SS
- Hemoglobin SC
- Sickle Beta-Plus Thalassemia
- Sickle Beta-Zero Thalassemia

How are the different common types of sickle cell disease inherited?
- Hemoglobin SS – The hemoglobin S gene is inherited from both parents
- Hemoglobin SC – The hemoglobin S gene is inherited from one parent and the hemoglobin C gene is inherited from the other parent
- Sickle Beta-Plus Thalassemia or Beta-Zero Thalassemia – The hemoglobin S gene is inherited from one parent and the beta thalassemia gene is inherited from the other parent.

Is one type of sickle cell disease worse than any other?
- The severity of sickle cell disease can certainly vary from person to person. Generally, Hb SS and Hb Sickle Beta-Zero Thalassemia tend to be the most severe and have the most complications.
- Hemoglobin SC – tends to be less severe than the above types. Tend to have less frequent and less severe pain crisis.
- Sickle Beta-Plus Thalassemia – tends to be one of the least severe types in African Americans. Low rate of complications, but should still be followed and get recommended routine screening.
- Hemoglobin C disease – mild disease- can have episodes of joint pain, mild anemia and mild jaundice.
- Hemoglobin AS- sickle cell trait. This is not sickle cell disease. Cannot develop sickle cell disease, but can pass it on to offspring. Have some hemoglobin S, but mostly normal hemoglobin (hemoglobin A).

Some Sickle Cell Facts
- The sickle gene is caused by a mutation in the hemoglobin gene.
- Hemoglobin is part of the red blood cell that carries oxygen around the body.
- Hemoglobin A is normal hemoglobin.
- Hemoglobin S is an abnormal type of hemoglobin.
- Most people with sickle cell disease do not have normal hemoglobin (hemoglobin A).
- Hemoglobin AS is sickle cell trait. This is not sickle cell disease.
- Sickle cell disease is usually diagnosed on New Born Screening when a baby is born.
Appendix J.

Sickle Cell Disease and Trait
The Georgia Comprehensive Sickle Cell Center at Grady Health System, Atlanta GA
www.SCInfo.org

What is Sickle Cell Disease?
Sickle cell disease is a group of inherited red blood cell disorders. It is the most common genetic disease in the US.

Over 70,000 Americans have sickle cell disease.

Normal red blood cells are round like doughnuts, and they move through small blood tubes in the body to deliver oxygen. Sickle red blood cells become hard, sticky and shaped like sickles used to cut wheat. When these hard and pointed red cells go through the small blood tube, they clog the flow and break apart. This can cause pain, damage and a low blood count, or anemia.

What makes the red cell sickle?
There is a substance in the red cell called hemoglobin that carries oxygen inside the cell. One little change in this substance causes the hemoglobin to form long rods in the red cell, when it gives away oxygen. These rigid rods change the red cell into a sickle shape.

How do you get sickle cell anemia?
You inherit the abnormal hemoglobin from both parents who may be carriers with sickle cell trait or parents with sickle cell disease. You can not catch it. You are born with the sickle cell hemoglobin and it is present for life.

Is Sickle Cell only in African Americans?
Sickle cell is in many nationalities including African Americans, Arabs, Greeks, Italians, Latin Americans and people from India. All races should be screened for this hemoglobin at birth. In the US, 1 out of 10 African Americans have sickle cell trait and 1 out of 625 newborns have the disease.

How can I be Tested?
A simple blood test called the hemoglobin electrophoresis can be done by your doctor or local sickle cell foundation. This test will tell if you are a carrier of the sickle cell trait or if you have the disease.

Newborn Screening
Most States now perform the sickle cell test when babies are born. The simple blood test will detect sickle cell disease or sickle cell trait. Other types of traits that may be discovered include: Hemoglobin C trait, Hemoglobin E trait, Hemoglobin S beta - which indicates an alpha thalassemia trait.

What is sickle cell trait?
Sickle cell trait is a person who carries one sickle hemoglobin producing gene - inherited from their parents and one normal hemoglobin gene. Normal hemoglobin is called type A. Sickle hemoglobin called S. Sickle cell trait is the presence of hemoglobin AS on the hemoglobin electrophoresis. This will NOT cause sickle cell disease. Other hemoglobin traits common in the United States are AC and AB traits.

Are there different types of sickle cell disease?
There are three common types of sickle cell disease in the United States.
1. Hemoglobin SS or sickle cell anemia
2. Hemoglobin SC disease
3. Hemoglobin sickle beta-thalassemia

Each of these can cause sickle pain episodes and complications, but some are more common than others. All of these may also have an increase in fetal hemoglobin which can protect the red cell from sickling and help prevent complications. The medication hydroxyurea also increases fetal hemoglobin.

Where can I get more information?
Visit the Sickle Cell Information Center on the internet at www.SCInfo.org or call the following:
The Georgia Comprehensive Sickle Cell Center at Grady Health System 404-616-3572
The Sickle Cell Foundation of Georgia Inc. 404-755-1641
Sickle Cell Disease Association of America 1-800-421-8453
REFERENCES


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VITA

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