

A Qualitative Exploration of the Experience of Adolescents and Young  
Adults and Parents in the Decision-making Process when Seeking  
Emergency Department Care for a Pain Episode in Sickle Cell Disease

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## Table of Contents

Acknowledgements.....	iii
List of Tables.....	vi
List of Figures.....	vi
List of Appendixes.....	vi
Abstract.....	vii
CHAPTER I.....	1
Introduction.....	1
Self-care and Disease Management in Adolescents/young adults.....	2
Healthcare Decision Daking in Adolescents/young adults.....	3
Purpose.....	5
Significance.....	5
Conceptual Framework.....	7
Study Aims.....	8
Conclusion.....	9
Overview of Chapters.....	9
Chapter II.....	17
Introduction.....	17
Aims.....	18
Methods/Methodology.....	19
Design.....	19
Search Methods.....	19
Search Outcome.....	20
Quality Appraisal.....	21
Data Abstraction.....	21
Synthesis.....	21
Results.....	22
Discussion.....	25
Conclusions.....	27
Literature Limitations.....	28
Chapter III.....	40
Specific Aims.....	40
Background.....	42
Sickle Cell Disease and Pain.....	42
Decision Making.....	44
Decision Making by Adolescents.....	45
Dialogical Self Theory.....	50
Part of Self or “me”.....	51
Adolescent and Young Adult.....	51
Culture.....	54
Experiences.....	57
Belongs to Me or “mine”.....	57

My Body.....	58
My Parent/Legal Guardian.....	59
My Doctor.....	61
Conclusions .....	61
Methods.....	62
Research Design.....	62
Population and Sample.....	63
Inclusion / Exclusion Criteria.....	63
Recruitment.....	64
Data Collection.....	67
Demographics.....	67
Qualitative Interviews.....	67
Data Management.....	69
Data Analysis.....	69
Limitations.....	75
Ethical Considerations.....	76
Institutional Review Board.....	76
Consent and Assent.....	76
Human Subjects.....	77
Protected Health Information.....	77
Potential Benefits.....	77
Data and Safety Monitoring Plan.....	78
Withdrawal from Study.....	78
Compensation.....	78
Chapter IV.....	98
Methods/Methodology.....	100
Design.....	100
Theoretical Framework.....	100
Study Setting and Recruitment.....	101
Inclusion and/or Exclusion Criteria.....	102
Data Collection and Ethical Considerations.....	102
Data Analysis.....	103
Findings.....	104
Discussion.....	109
Limitations.....	112
Recommendations for Further Research.....	113
Conclusions.....	114
Chapter V.....	149

### List of Tables

Table 1	Study Characteristics Quantitative.....	30
Table 2	Study Characteristics Qualitative.....	31
Table 3	Parent/Legal Guardian Demographics .....	122
Table 4	Adolescent and Young Adult Demographics .....	122
Table 5	Quotes by Adolescents/young Adults .....	123
Table 6	Quotes by Parents and Legal Guardians.....	126

### List of Figures

Figure 1	Analysis using Dialogical Self Theory.....	11
Figure 2	Search Flowsheet.....	29
Figure 3	Dialogical Self Theory.....	80
Figure 4	Dialogical Self Theory .....	115
Figure 5	Theme Development .....	116

### List of Appendixes

Appendix A	Semi-structured Interview Guide Adolescents.....	81
Appendix B	Semi-structured Interview Guide Parents.....	83
Appendix C	Phone Script.....	117
Appendix D	Semi-structured Interview Guide Adolescents.....	118
Appendix E	Semi-structured Interview Guide Parents.....	120

### **Abstract**

Sickle cell disease affects individuals in the United States and worldwide. It is an inherited genetic disorder that is marked by recurrent pain episodes as well as organ damage. Parents/legal guardians are legally responsible for making medical decisions for adolescents under the age of 18 years. However, once an adolescent becomes an adult, they are responsible for making medical decisions. The purpose of this study was to explore the thought process of adolescents and young adults with sickle cell disease, as well as their parents/legal guardians in healthcare decision making when considering emergency department treatment for acute vaso-occlusive pain episodes. This study used qualitative descriptive methods with thematic analysis. Nine parents/legal guardians participated in the study; six adolescents under the age of 18 years and one young adult aged 19 agreed to participate for a total of 16 participants. Four themes emerged from the analysis describing the experiences of the participants healthcare decision making regarding whether to use emergency department care: (1) Knowledge acquisition through learning activities; (2) Thought process in healthcare decision making; (3) Discussion to seek vaso-occlusive episode care in the emergency room through consideration of reasons for emergency department care; and (4) Reasons it is the responsibility of parents/legal guardians of children. As adolescents and young adults transition to adult care they become the primary decision-maker for their healthcare needs. It is important that parents/legal guardians and healthcare providers ensure there is an understanding of the process of healthcare decision making prior to transition.

Key words: Sickle cell disease, healthcare decision making, adolescents and young adults, pain episode

## Chapter I

### Introduction

Sickle cell disease (SCD) affects approximately 100,000 individuals in the United States. Approximately 1 of every 365 Black or African American infants and 1 of every 16,300 Hispanic American infants are born each year in the United States with SCD (CDC, 2022). Sickle cell disease is an inherited genetic disorder that is marked by recurrent pain episodes as well as organ damage caused by vaso-occlusion (Ware et al., 2017). Four phases of pain have been identified in vaso-occlusive episodes (VOE). The prodromal or phase I is marked by numbness, aches, and paresthesia with the beginnings of inflammation and is present prior at onset of VOE pain. In phase II or the initial phase pain is established and becomes severe, reaching a higher level. It is at this higher level of pain that treatment is recommended (Ballas, Gupta, & Adams-Graves, 2012; Jacob et al., 2005). During the third or established phase pain is severe and steady lasting approximately three to seven days and is often marked by hospital admission. The fourth or resolving phase lasts eight to ten days during which time there is a gradual decrease in pain intensity and patients are often treated at home (Ballas et al., 2012; Jacob et al., 2005). Parents/legal guardians are legally responsible for making medical decisions for adolescents under the age of 18 years. Despite recurrent VOEs in this population, it is unknown what level of adolescent involvement is included in the healthcare decision making (HCDM) processes surrounding SCD. An adolescent, defined as being between 14 and <18 years, is not generally considered capable of making medical decisions on their own (Hartman, 2002).

Treatment for VOE begins at home with increased water consumption, topical heat, over-the-counter analgesics, and opioids (Brandow et al., 2020). Vaso-occlusive episodes

that do not respond to home care and progress to phase II require emergency medical treatment with intravenous fluids, oxygen, and intravenous or nasal opioids. The process of seeking treatment requires HCDM by the parent/legal guardian and/or the adolescent/young adult (AYA). It is unknown how HCDM occurs when an AYA requires emergency department care.

### ***Self-care and disease management in adolescents/young adults***

Adolescents are defined as age 14 to <18 years old. Young adults are typically defined as individuals aged 18 to 21 years old. This population can experience a variety of chronic diseases that require daily decisions to be made in daily management. These decisions may not always be in collaboration with the parent/legal guardian (Miller & Drotar, 2007). Self-care and disease management for AYAs has been studied in the literature, especially for those who have diabetes. Diabetes requires daily HCDM involving testing of blood sugar, taking medications, diet, and exercise (American Diabetes Association, 2021).

Parent-adolescent communication in daily decision-making has been addressed in studies focusing on the treatment of Type I diabetes. One study focused on 62 parent-adolescent dyads (Miller & Drotar, 2007). Parents indicated that higher negative communication by the adolescent resulted in lower adolescent adherence to care ( $r = -.28$ ,  $p < .03$ ), and providers reported lower adherence by adolescents ( $r = -.26$ ,  $p < .04$ ). Children and AYAs with a range of chronic diseases have expressed varying degrees of interest in decision-making participation (Coyne, Amory, Kiernan, & Gibson, 2014; Vaknin & Zisk-Rony, 2011). Most who wanted involvement in decision-making expressed a need to have physician and parent input (Garth, Murphy, & Reddihough, 2009; Jedeloo, van Staa,

Latour, & van Exel, 2010). Chronic disease may have a negative impact on adolescent development, resulting in a delay of said development (Holmbeck, 2002). Based on this research, the best way for an adolescent to learn self-care decision making skills may be through collaboration with a parent/legal guardian. It is unknown if and at what age this should start. Further research is needed to assess how parent/adolescent collaboration develops and affects decisions made for the child.

Another chronic disease requiring self-care management is AYAs with SCD. Unfortunately, there is sparse literature regarding decision-making in SCD. Varty et al. (2022) explored decision-making in 50 AYAs with SCD. It was found that AYAs demonstrated a higher level of health care responsibility if they participated in shared HCDM. Odityo (2023) explored AYA perceived involvement in shared decision making related to SCD self-care management and health outcomes with 24 AYAs responding to an email. It was noted that ability and actions related to self-care were significantly related to perceived shared decision making. These findings indicate that more research is needed related to parent/adolescent communication regarding chronic illness to ascertain the best way for parents to interact with their adolescents. It also indicates that parents may need to start conversations about self-care prior to adolescence to prevent some negative care discussions and increase adolescent autonomy.

### ***Healthcare decision making in adolescents/young adults***

Cognitive development is critical when determining decision-making ability. Individuals with SCD may have impaired cognitive development due to recurrent or chronic low hemoglobin and hematocrit levels, overt stroke, silent stroke, or other reasons related to SCD (Prussien et al., 2020; Schatz, Finke, Kellett, & Kramer, 2002). There is a

positive correlation between cognitive ability and good decision-making (Parker & Fischhoff, 2005).

Vaso-occlusive episodes are the most common cause of hospitalization and emergency department visits in SCD (Brousseau, Owens, Mosso, Panepinto, & Steiner, 2010). Collaborative decision-making for treatment of VOE pain involves the parent/legal guardian and AYA, taking into account activities such as AYA advice-seeking and parent/legal guardian and AYA negotiation (Miller, 2009). Adolescents and young adults tend to seek adult input less often as they age and rely on past experience or peers to assist with HCDM (Byrnes, 2002). Not all parents allow the adolescent to collaborate in the HCDM process, despite the adolescent being developmentally ready to participate (Miller & Drotar, 2007). Translating knowledge from parents/legal guardians into usable behaviors is dependent on AYA cognitive ability. Knowledge from research and healthcare provider teaching can be translated into workable behaviors (Straus, Tetroe, & Graham, 2011), thus the healthcare provider should have taught the parent from the time of diagnosis how to care for and make decisions about healthcare for the child.

Different experiences and perceptions are based on the role of participant in the phenomena being studied (Patton, 2002). Thus, a parent/legal guardian and an AYA may perceive the collaborative process of HCDM in different ways, describing reality and experiences from two very subjective perspectives. While both perspectives are real, they do not indicate that one is more true or important than the other. Each participant has constructed their reality based on their personal experiences and implications of the phenomena may have on their individual lives. A cross-sectional, correlational study by Varty et al. (2022) of 50 AYAs with SCD and their caregiver administered the Decision-

Making Involvement Scale. The purpose of the study included exploration of decision-making involvement in healthcare responsibility. Adolescents and young adults reported that participation in discussion trading decision-making healthcare was associated with higher levels of healthcare responsibility (Varty et al., 2022). While there is some research regarding HCDM in chronic disease, this work is all but non-existent in SCD, a chronic disease that requires frequent life-impacting decisions regarding care seeking and treatment adherence.

## **Purpose**

The purpose of this study was to explore the thought process of adolescents and young adults with sickle cell disease, as well as their parents/legal guardians in healthcare decision making when considering emergency department treatment for acute vaso-occlusive pain episodes. This study explored 1) the thought processes of HCDM by AYAs with SCD, aged 14 to 21 years, and their parents/legal guardians when considering seeking emergency department care for VOE and 2) the perception of the readiness of the AYA to assume HCDM by AYAs and parents/legal guardians. Factors evaluated by AYA, and parent/legal guardian may differ between AYAs and parents/legal guardians, and parents/legal guardians' beliefs that AYAs are ready to begin with the assistance of HCDM may vary. This research employed descriptive qualitative methodology to examine the phenomenon of AYAs making decisions to seek emergency department care for VOEs (Braun & Clarke, 2006).

## **Significance**

Sickle cell disease is a life-long disease with the prevalent complication of VOE beginning in infancy with dactylitis or pain in the hands (Ware et al., 2017). Vaso-occlusive

episodes continue throughout life, often increasing in frequency and severity, beginning in the teenage years and presenting in the long bones of the extremities. While VOE pain often subsides after treatment, there is a possibility of mortality due to frequent episodes of VOE or other complications of SCD. Mortality rates in AYAs with SCD increase between ages 15 and 19 years and further increased between 20 and 24 years of age (Piel, Steinberg, & Rees, 2017). As AYAs age, their healthcare utilization for SCD complications increases, being most prevalent between the ages of 18 and 30 years (Brousseau, Owens, et al., 2010; Ware et al., 2017), thus requiring HCDM regarding type of care and where to seek treatment.

During adolescent development, health habits and trajectory can be positively or negatively developed. While adherence to treatment may be negatively affected, problem solving may positively improve (Holmbeck, 2002). As adolescents age, they develop autonomous health behavior (Wray-Lake et al., 2010). Adolescents with chronic diseases take into account the role they feel they have in HCDM as well as that of the parent while considering their current quality of life regarding their health (Lipstein, Muething, Dodds, & Britto, 2013).

Adolescents and young adults experiencing a variety of chronic illnesses are required to make daily decisions in the management of their disease. While decisions may not always be in collaboration with the AYA, decisions to go to the emergency department for care are generally made by the parent/legal guardian and/or the young adult aged 18 or older. The HCDM process is not well understood in the AYA population with SCD. It is also unknown how involved AYAs who live with or are in close contact with their parent/legal guardian when caring for the SCD make healthcare decisions. Given that there

are many factors to consider during HCDM, it is imperative that AYAs appropriately consider past responses to emergency department treatment, implications of not seeking emergency department care, and other factors related to their health and experiences.

### **Conceptual Framework**

Decision-making capacity is defined by the processing of information and comprehension by the AYA with an understanding of the disease and proposed treatment information provided by the healthcare (Ruhe, Wangmo, Badarau, Elger, & Niggli, 2015). A decision takes into account available treatment options and behaviors related to selection of the option believed to provide the desired outcome (Byrnes, 2002). There are many factors to be considered in HCDM. Healthcare decision-making capacity considers the difficulty of decision and requires the AYA to clearly communicate their decision. Capacity varies between small choices and more important decisions and may vary depending on experience with HCDM and maturation level.

Dialogical Self Theory was used in this study to examine internal conversations related to the HCDM process. Exploration of this process related to visiting the emergency department for a VOE due to SCD aided in understanding how AYAs processed thoughts related to the HCDM process and indicated needed teaching prior to transition to adult care.

Dialogical Self Theory was used to explain the self in relation to the thought process with the HCDM process (Figure 1). The self is composed of two parts: one part of the self is located within the mind and the other part extends to the environment (Hermans, 2012). Dialogical Self Theory was used in this study to analyze internal conversations related to the HCDM process in AYA with SCD.

Mikhail Bakhtin added the concept of the involvement of multiple voices to inner dialogue as cited in Hermans (2001). These multiple voices are portrayed as thoughts within the mind of the person making the decision and may potentially include parents/legal guardians, physicians, and community. Collective voices or thoughts are influenced by health care providers, jargon, culture, social circles, teachings, and other such entities. As a result of this community of voices, the decision makers may have conflicting thoughts about their decisions. Further, thoughts about healthcare providers and parent/legal guardian taught and treatment within the hospital may work to dominate the discussion with the parent/legal guardian. Exploration of the influences of these factors is needed to understand the AYA thought process that results in positive methods of teaching and addressing healthcare needs, noting that the experiences become part of the self. These voices, both as part of the self and as belonging to me, i.e. the AYA, become a story, agreeing, and disagreeing, that results in the development of a solution to the discussion at hand based on possessed knowledge and experience. Exploration of this process related to visiting the emergency department for a VOE due to SCD aided in understanding how AYAs process thoughts related to HCDM and indicate needed teaching prior to transition to adult care.

### **Study Aims**

**Aim 1.** Identify and compare factors that influence the parent/legal guardian's and AYA's healthcare decision-making when deciding to seek care from the emergency department for VOE in SCD.

**Aim 2.** Compare and contrast parent/legal guardian and AYA decisions about the AYA's capabilities of seeking emergency department care for VOE in SCD.

## **Conclusion**

Understanding the definitions and complex interplay of concepts related to HCDM in AYAs is critical to the development of AYA autonomy and clinical interventions that address these dynamics. Describing concepts clearly and understandably will also facilitate the dissemination and use of these research findings, making them accessible to experts outside the specialty of SCD management. More research is needed related to parent/adolescent communication regarding chronic illness to ascertain the best way for parents to interact with their adolescents. It also indicates that parents may need to start conversations about self-care prior to adolescence to prevent some negative care discussions and increase adolescent autonomy.

## **Overview of Chapters**

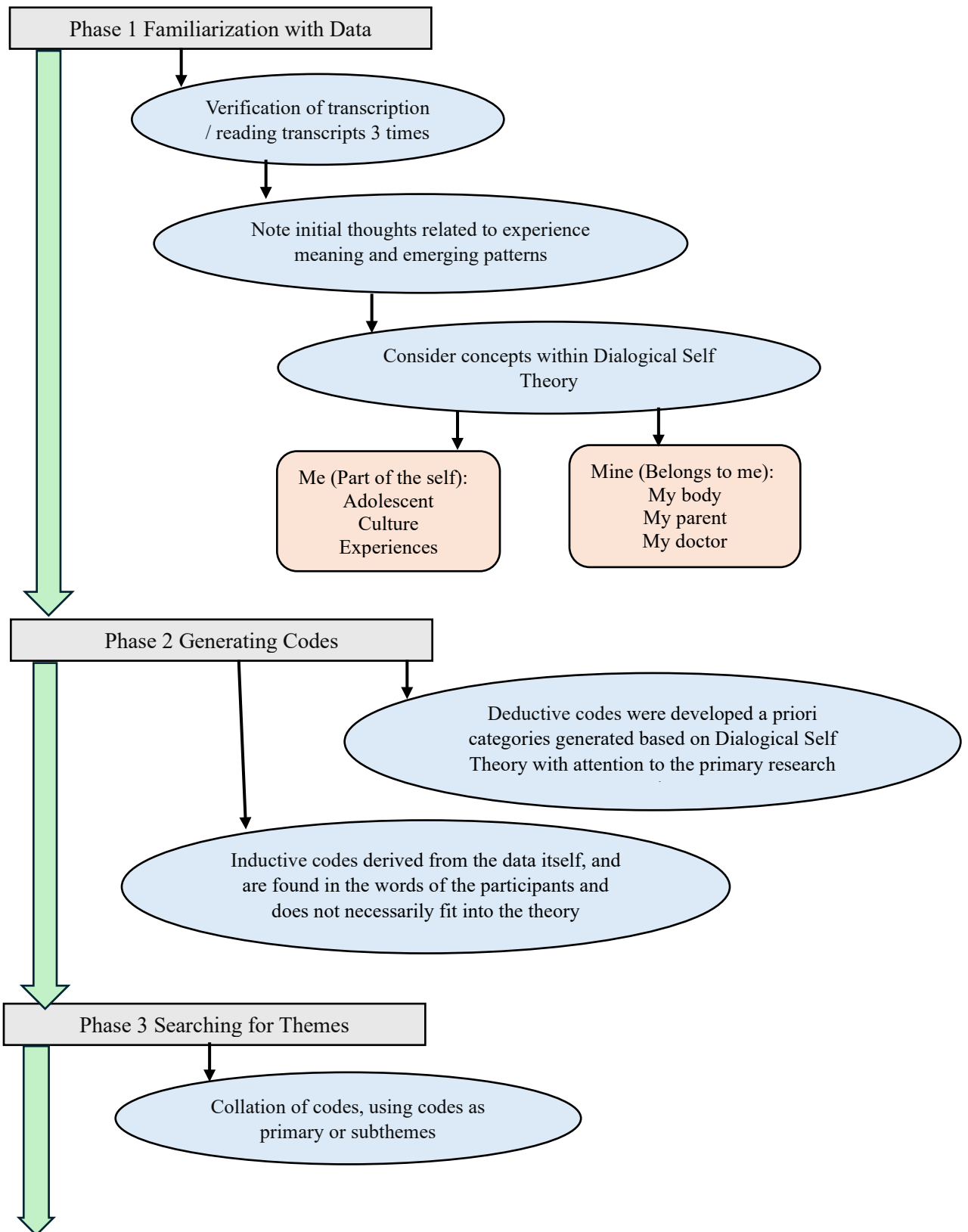
Chapter II is a systematic literature review related to AYA HCDM regarding chronic disease, transition preparation to adult care and Dialogical Self Theory. Results of the review indicate that AYAs want to participate in HCDM in an active or shared approach. Factors influencing HCDM include quality of life, health issues, and social interactions. Recommendations include more research to explore the benefits of education in the HCDM process to improve healthcare management of AYA seeking emergency department care for acute VOE. The review used the Preferred Reporting Items for Systematic reviews and Meta-Analyses (PRISMA) guidelines.

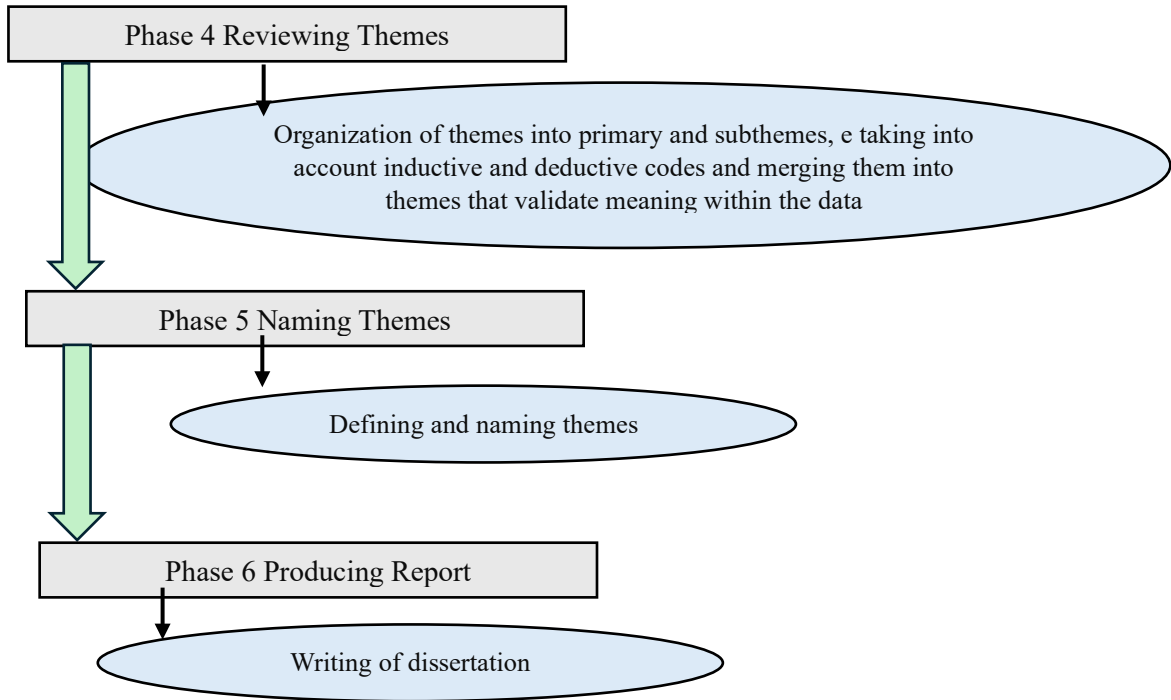
Chapter III is the approved research proposal. Methods are described in detail, including the study's theoretical underpinnings, timelines, participants and recruiting, ethical considerations, data collection, and data analysis.

Chapter IV discusses key findings from the study. Study conclusions, and implications of the findings for future teaching needs in the SCD AYA transition program to adult care are discussed.

Chapter V discusses the study results and implications of the findings for future research and clinical practice.

Figure 1 Analysis using Dialogical Self Theory





(Braun & Clarke, 2006)

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## Chapter II

### Introduction

Approximately 4.5 million or 18.4% of adolescents aged 12 to 18 years have a chronic healthcare condition in the United States (McManus et al., 2013). Adolescents and young adults (AYAs) with chronic disease transition to adult care between the ages of 18 and 21 years, depending on site-specific clinic practice. While chronic disease treatment is much improved and AYAs more frequently live to adulthood, there are many issues related to transition to adult care that need addressed. One issue is that AYAs experiencing a variety of chronic diseases require daily healthcare decision-making (HCDM) skills to manage their disease as well as diagnostic and treatment decision-making abilities. Decisions take into account available options resulting in selection of a treatment believed to provide the desired outcome (Byrnes, 2002). Thus, HCDM is important to health maintenance as AYAs become independently responsible for their care.

Decision-making capacity is defined as the processing of information and comprehension by the individual with an understanding of the disease and proposed treatment information provided by the healthcare provider (Ruhe et al., 2015). Healthcare decision making capacity considers the difficulty of the decision and requires an individual to clearly communicate their decision. Decision-making capacity varies between small choices and more important decisions and may vary depending on experience with HCDM and the individual's maturation level.

Parents/legal guardians are legally responsible for making healthcare decisions for adolescents. An adolescent, per United States law, is defined as being between 14 and <18 years, and is not generally considered capable of making healthcare decisions independently (Hartman, 2002). However, once an adolescent is considered an adult (18

years or older), they are responsible for making independent healthcare decisions. Autonomous self-care skills should evolve to become an independent activity as the AYA transitions to adult health care (Castillo & Kitsos, 2017). This is especially important with chronic diseases, as disease may include frequent occurrences of complications and the need to understand the impact of such complications on the body and resultant treatment needs. Thus, autonomous HCDM should evolve to become an independent activity as the AYA transitions to adult health care.

Healthcare decision making is becoming an increasingly studied field with AYA inclusion in their treatment and care planning. Autonomy-seeking in adolescence is an activity that impacts independence (Spear & Kulbok, 2004). While it is not known how or whether AYAs may be involved in decision-making, it is known that not all parents/legal guardians allow the adolescent to participate in the decision-making process, despite the adolescent being developmentally ready to participate (Miller & Drotar, 2007). This review seeks to explore HCDM in AYAs within the pediatric care setting regarding care and treatment of chronic disease.

### **Aims**

The aim of this systematic review is to synthesize existing knowledge of AYAs with chronic disease regarding involvement in HCDM, their thought process in decision-making, and the way they make decisions with or without their parent/legal guardian. This review will: (1) identify studies in which AYA participation in HCDM was explored and described; (2) identify concepts from included papers relevant to AYAs desire to participate in HCDM and the decision-making process utilized; and (3) identify types of

parent/legal guardian and healthcare provider support desired by the AYA decision-making.

## **Methods/Methodology**

### ***Design***

A systematic review of mixed methods literature was conducted. The use of this method confirms that all relevant evidence was examined, and that data were synthesized to generate valid findings. The Preferred Reporting Items for Systematic Reviews and Meta-Analysis (PRISMA) guidelines (Moher, Liberati, Tetzlaff, Altman, & PRISMA Group\*, 2009; Page et al., 2021) was followed, including the identification of inclusion and exclusion criteria, essential evaluation of included studies, and inclusion of qualitative and quantitative findings. The literature search was completed in October 2023.

### ***Search Methods***

A research librarian was consulted, and databases searched for articles in English included CINAHL, Scopus, Google Scholar, Pub Med and EBSCO Open Dissertations. Key words used were “clinical or healthcare decision-making”, “chronic disease or illness”, “adolescents or young adults” to include years 2013 through 2023 and full articles or conferences. Articles were reviewed to include qualitative and quantitative studies. The search was expanded by looking at references of pertinent studies and author names and Cochrane Reviews.

Inclusion and exclusion criteria used in the review includes only empirical research on AYA participation in HCDM in the context of chronic disease or illness related to HCDM for treatment, diagnostics, daily management, and healthcare interventions, eliminating decision-making regarding research study participation. Articles outside of the

United States were excluded due to possible cultural differences related to HCDM in the clinical setting. Literature regarding reproductive health and fertility in cancer survivorship was excluded because it does not directly relate to disease management. Studies related to HCDM about mental health were excluded because not all AYAs may be capable of developing autonomous decision-making skills, depending on diagnosis and severity of mental illness. Literature regarding decision aids was excluded because this systematic review focused on the decision-making process without the assistance of decision-making tools. Studies which included children with a mean age below 13 years or adults with a mean age greater than 21 years were also excluded.

Data were extracted from eligible studies based on the purpose of this paper to include HCDM involvement by AYAs, their thought process in decision-making to include factors considered, and the way they make decisions with or without their parent/legal guardian. Data from the AYA included age, disease, and treatment decision. Data further included AYA type of decision making in qualitative studies and study purpose in quantitative studies (there were no intervention studies identified). Data on outcomes included involvement of AYAs in HCDM and consideration of factors affecting decision-making. Study titles and abstracts were reviewed to determine relevance and for inclusion/exclusion criteria.

### **Search Outcome**

The study selection progression is summarized in Figure 2. There were 21,321 articles identified during the initial literature search. Duplicates and articles that noted exclusion criteria based on the title were excluded. Fifty-four articles were then assessed for eligibility and read in full. Twelve articles were selected for inclusion in the analysis.

### **Quality Appraisal**

All articles were reviewed for quality assessment by the first author (DR) as described by Kmet, Cook, and Lee (2004). The first author evaluated and scored all articles, noting that it was possible to score up to 20 qualitative studies and 28 quantitative studies. Scoring criteria included such things as study design appropriateness, question/objectives well described, appropriate sample size, appropriate analysis described, and conclusions supported by the results. Scores ranged from 16-19 for quantitative studies n=4 (none of the studies were interventional thus final scores were lower), and 12-19 for qualitative studies n=8 All articles were included in this review.

### **Data Abstraction**

Data were extracted from the reviewed articles (Table 1 and Table 2). Headlines used for quantitative design included author/year/chronic illness, aims, study design, sample, and outcomes. Those used for qualitative design included author/year/chronic illness, aims, sample, type of decision-making explored, outcomes.

### **Synthesis**

Qualitative articles were analyzed using inductive content analysis methods, adding words and segments into codes and then developing them into main concepts (Elo & Kyngäs, 2008) using MAXQDA 2022. Each article was reviewed four times to assign codes. Codes were then grouped into concepts such as desire to participate in HCDM by AYA and factors considered by AYA when considering treatment plan. Quantitative articles were analyzed using narrative synthesis (Popay et al., 2006) to combine findings from multiple studies and combine them into a textual summary of written material, leading

to the identification of data related to study design, study purpose, findings, and implications. The data were entered into an excel spreadsheet to perform this analysis.

## **Results**

Eight qualitative studies focusing on HCDM in AYAs were found. Qualitative study participants were AYAs ages 12 to 25 years except two studies: one included adults to age 39 years McGrady, Brown, and Pai (2016); and Fremont, Slick, Willi, Miller, and care (2021), who included 8 through 17 year olds with an average age of 13.7. Chronic diseases included cardiac issues, rheumatologic illnesses, cancer, sickle cell disease and hypercholesterolemia. There were three main concepts associated with the findings of all qualitative articles in this review: 1) AYA wanted to participate in HCDM, noting different types of desired participation, 2) factors considered by AYAs when making decisions, and 3) factors considered by parent/legal guardians when making decisions. There were three main concepts associated with the findings of all qualitative articles in this review: 1) AYA wanted to participate in HCDM, noting different types of desired participation, 2) factors considered by AYAs when making decisions, and 3) factors considered by parent/legal guardians when making decisions.

Three cross-sectional and one correlational study focusing on HCDM by AYAs in chronic disease treatment. Participants were aged 12 to 25 years old with an average range of 16 to 18 years old. Chronic diseases included cancer, SCD, diabetes, Crohn's Disease, and juvenile idiopathic arthritis. All studies found that AYAs wanted to and/or did participate in HCDM. The AYAs also wanted to participate in a collaborative process of HCDM to include discussion with their parents and/or healthcare provider.

Results indicated that AYAs desired to and/or participated in HCDM, with most desiring active or shared participation (Fremont, Slick, Willi, Miller, et al., 2021; Lipstein et al., 2013; Pyke-Grimm, Franck, Halpern-Felsher, Goldsby, & Rehm, 2022; Sliwinski et al., 2017; Weaver et al., 2015). Some AYAs desired input based on factors such as treatment experience, autonomy, personal and parent knowledge of illness and treatment, and knowing that preferences would be honored (Lipstein et al., 2013; Sliwinski et al., 2017; Weaver et al., 2015). Trust by parents/legal guardians and healthcare providers in their ability to take responsibility for their health was a factor in AYA desire to become the primary decision-maker (Fremont, Slick, Willi, & Miller, 2021). Adolescents and young adults desired inclusion in the discussion of their preferences and their knowledge of their health and the treatment plan (Fremont, Slick, Willi, & Miller, 2021; Weaver et al., 2015). Further, they wanted support in their decision making role from their healthcare providers (Fremont, Slick, Willi, & Miller, 2021; Weaver et al., 2015). Ability to comply, defined as a decision to take responsibility for treatment follow-through was found to be influenced by education regarding the prescribed treatment (Pyke-Grimm et al., 2022). Adolescents and young adults noted that becoming self-aware of the need to begin HCDM involved developing autonomy, managing treatment, and finding credible sources for guidance. Autonomy included taking responsibility for health care needs. Other factors included adverse effects related to treatment; impact of treatment modalities, and time management on quality of life to include leisure, family and school activities, discomfort of treatment, and impact on family members; and expected outcomes and prognosis (Lipstein et al., 2013; Lipstein, Dodds, Lovell, Denson, & Britto, 2016; McGrady et al., 2016; Needle et al., 2020). Parents/legal guardians considered potential side effects, disease characteristics,

knowledge based on personal research, treatment risks and benefits and AYA quality of life, to include interference of daily activities (David, Lo, & Langer, 2018; Lipstein et al., 2013; Lipstein et al., 2016).

Eight qualitative studies focusing on HCDM in AYAs were found. Qualitative study participants were AYAs ages 12 to 25 years except two studies: one included adults to age 39 years McGrady et al. (2016); and Fremont, Slick, Willi, Miller, et al. (2021), who included 8 through 17 year olds with an average age of 13.7. Chronic illnesses included cardiac issues, rheumatologic illnesses, cancer, sickle cell disease and hypercholesterolemia. Recruitment occurred in the outpatient clinical setting or the local hospital.

Four quantitative studies were reviewed, two studies with parent/legal guardian and AYA dyads while the other two included only AYAs. Three studies used cross-sectional design and the fourth was a correlational study. Three studies explored aspects of AYA involvement in HCDM and reported the majority of AYAs preferred to be actively involved (Cousino et al., 2020; David et al., 2018; Miano et al., 2020). Cousino et al. (2020) explored preferences for an AYA treatment plan. Of 53 AYAs, 45.3% expressed a desire to actively participate in patient led HCDM regarding treatment and 35.3% of 51 AYAs preferred shared decision-making with the physician. They further noted that AYAs desired active involvement in discussion surrounding factors such as adverse effects or treatment risks and prognosis when considering treatment options. The longer the AYA experienced the heart condition, the more involved they wanted to become. Miano et al. (2020) found that 39% (n=46) of AYAs wanted to actively participate and make the final decision while 34% wanted to be involved through shared decision-making with the

provider. There was a positive correlation between perceived involvement of AYAs in HCDM and desired involvement (n=28) with less decisional conflict when increased participation was noted (David et al., 2018). Varty et al. (2022) explored the relationship between decision-making involvement and health-care responsibility, noting higher levels of health care responsibility were associated with sharing opinions and ideas in decision-making (n=50).

### **Discussion**

This mixed methods systematic review demonstrates a desire by AYAs to actively or in a shared decision-making capacity with parents/legal guardians and healthcare providers participate in HCDM. Active and shared decision making were the primary means of involvement identified as being desired by AYAs. Two studies in this review found that AYAs wanted to involve parents/legal guardians and/or the healthcare provider as partners in shared decision making (Sliwinski et al., 2017; Weaver et al., 2015).

Shared decision making involves a minimum of two individuals (Charles, Gafni, & Whelan, 1997), generally the patient and the healthcare provider, but in the case of AYAs a third individual, the parent/legal guardian may also be involved. Shared decision making requires discussion, acknowledgement of expectations and harmonizing of roles between the participants so a decision around the healthcare issue being discussed may be reached (Charles et al., 1997). Discussion between a healthcare provider and AYA provides the AYA with a better understanding of the proposed plan of care, including its purpose, procedures involved, possible side effects and risks, alternative options, and expected outcome (Halpern-Felsher & Cauffman, 2001). The healthcare provider has an ethical responsibility to assure such information is shared with and understood by the AYA so

they may express their preferences through open discussion and participate in informed decision-making (Coyne et al., 2014; O'Hair et al., 2003).

This review found that AYAs had a desire to participate in HCDM activities, thus indicating a need for healthcare providers and parents/legal guardians to assure participation occurs at a level desired by the AYA. Transition programs require provision of adequate information be given to AYAs to assure their knowledge and understanding of their chronic disease and thus enable participation in HCDM (Willis, McDonagh, & Practice, 2018). Adult care requires patient autonomy and independence by the young adult in self-care and decision making,(Castillo & Kitsos, 2017), thus requiring transition planning to be focused on these needs.

Autonomy is a key factor of development as AYAs mature and grow cognitively and emotionally (Spear & Kulbok, 2004). As AYAs develop, they become independent in caring for themselves emotionally, socially, and physically (Melendro, Campos, Rodríguez-Bravo, & Arroyo Resino, 2020). These changes lead to the development of independent decision-making skills in healthcare. Their capacity to make decisions includes the ability to contemplate treatment options based on research, discussion, and understanding of information (Melendro et al., 2020).

Parents/legal guardians decision making process for their children has focused on whether disease would worsen without treatment, treatment benefits, and possible side effects, (Allen, 2014; Lipstein et al., 2016). Other factors considered by parents/legal guardians included disease characteristics related to symptoms (Lipstein et al., 2016). Risks and benefits were considered by both AYAs and parents/legal guardians, but were explicitly defined by parents/legal guardians (Lipstein et al., 2016). Parents/legal guardians

wanted specifics regarding disease severity, to include such things as chromosomal abnormalities and structural defects. They also wanted to understand treatment efficacy and prognosis. Healthcare providers' demeanor and presentation were influential in HCDM, and parents/legal guardians wanted to experience a sense of hope. Other factors included spiritual and religious beliefs (Allen, 2014). This review found limited understanding of these factors influencing AYA HCDM.

Overall, this study shows that AYAs have a desire to participate in HCDM but are not well equipped to do so. While parents/legal guardians of children with chronic disease evaluate important factors related to the current health and expected impact of treatments being offered, this research suggests that AYAs need education regarding factor evaluation. Parents/legal guardians report that decision-making skills evolve over time with treatment being the most frequent consideration (Lipstein & Britto, 2015). Autonomous decisions require adequate knowledge of disease and treatment options (Kukla, 2007). Based on findings, learning about disease, health care and treatment options before requiring decisions to be made should be overseen by parents/legal guardians and healthcare providers. Findings also suggest that parents/legal guardians and healthcare providers should include adolescents in the decision-making process at an early age to allow for the evolution of the decision-making process.

## **Conclusions**

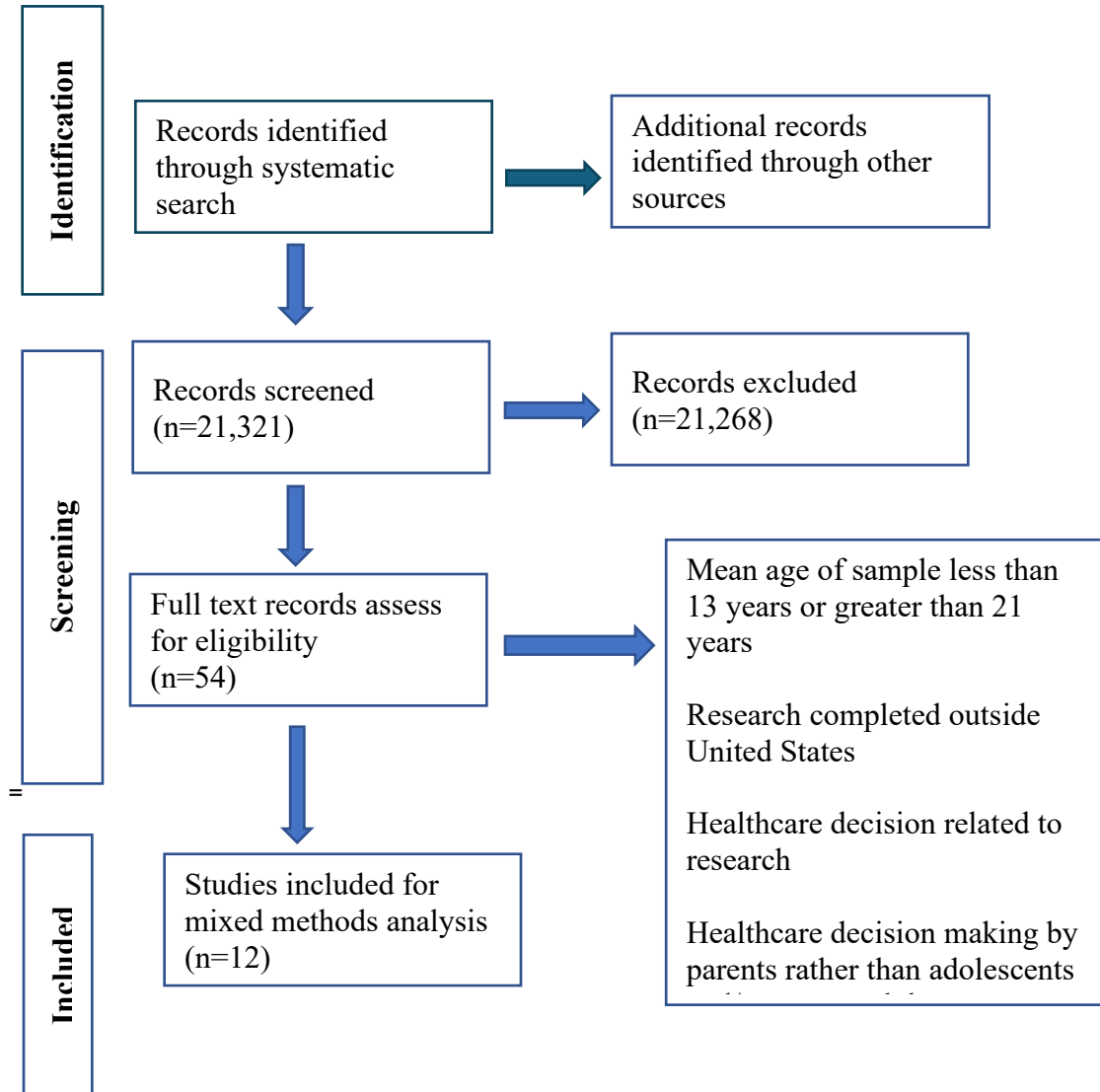
Decisions such as whether to use medication, when to seek emergency medical care, and what treatments are needed vary among those with asthma, cancer, diabetes, multiple sclerosis, sickle cell disease and other chronic diseases. This review suggests that the process of decision-making includes four factors which were identified as 1) health

implications, 2) past experiences, 3) teaching, and 4) parent input needs further exploration. One of the factors, teaching, is an essential factor to address and consideration should be explored to evaluate AYA abilities to make appropriate decisions for both short-term and long-term medical care. Examining the process of HCDM by AYAs and developing a program of education to assure that decisions are of high quality and results will meet the needs of the AYAs as a prerequisite when preparing AYAs for transition to adult care and independence as well as shared decision making with healthcare providers is crucial because it will empower the young adult to take ownership of their health which will ensure continuity of care and managing chronic conditions effectively ultimately promoting better health outcomes in the long-term.

### **Literature limitations**

There are a few limitations to this review. One limitation is that while these studies demonstrate a desire for AYAs to participate in HCDM, studies that seek to understand factors considered by AYAs related to medical needs describe factors on a limited basis and do not explore explicit thought processes regarding impact on decision-making. A second limitation is a lack of randomized control trials that seek to understand the decision-making process of AYAs or the outcome of these decisions. Third, these studies consist of small samples. While this is expected in qualitative research, the lack of larger samples in quantitative studies limits abilities to apply findings to the chronic disease population. Finally, there is no research that explores parents or healthcare provider teaching as AYAs are preparing for transition to adult care.

Figure 2 Search flowchart  
 AYA participation in  
 Healthcare Decision



<b>Table 1</b>				
Study Characteristics Quantitative				
Author	Aims	Study Design	Sample	Outcomes
Year				
Chronic Illness				
Cousino, M.K., et al. 2023 Heart Transplant	Examination of decision-making inclinations and describe preferences for medical and end-of-life decision making	Cross-sectional study using survey method	12 AYAs, 12-24 years (M=15.2, SD=2.4)	82% reported involvement in decision to be put on transplant list via parent asking AYA for input while only 42% discussed end-of-life decision-making.
David, J.G., M.S. Lo, and D.A. Langer 2018 Chronic rheumatologic illnesses	To evaluate correlations of adolescent and parent medical decision-making involvement	Correlational study	28 adolescents (M=16.1 years, SD=2.5) and parent dyads	There was more decision-making involvement in adolescents who thought participation to be important and favored involvement. Less decisional conflict resulted in increased participation.
Miano, S.J., et al 2020 Cancer	Exploration of the amount of control adolescents and young adults prefer with complex medical decision making	Cross-sectional descriptive correlational design	32 AYAs 14-25 years (M=18.2, SD=3.2)	39% favored active collaboration while 34% wanted shared decision-making
Varty, M., et al. 2022 Sickle cell disease	Explore relationship between decision-	Cross-sectional correlational study	50 AYAs 13-21 years (M=17.12,	AYAs with higher levels of health care responsibility

making involvement and health care responsibility	SD-2.90) – parent dyads	were associated with sharing opinions and ideas in decision-making
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<b>Table 2</b>				
Study Characteristics Qualitative				
Author	Aims	Sample	Type of decision-making explored	Outcomes
Year				
Chronic illness				
Fremont, E.R., et al. 2021 Type 1 diabetes	Exploration of parent and youth perspectives of process of decision-making	41 children and their parent dyads 8-17 years (13.7 ± 2.9)	continuous glucose monitoring	The majority of youth and parents described youth as the primary decision maker (54.8%) while only a few expressed beliefs that parents should be primary caregivers (4.8%). It was found that Parents should provide input in the decision made by youth (14.3%).
Lipstein, E.A., et al. 2013 Crohn's Disease and juvenile idiopathic arthritis	Exploration of adolescent roles and preferences in HCDM	15 youth 12-17 years n=8 JID, n=7 CD)	TNF-xi	Many adolescents want to be involved in decision making about quality of life and disease treatment. They want parent and physician input.

Lipstein, E.A., et al. 2016 Crohn's Disease and juvenile idiopathic arthritis	Comparison of factors considered by parents to those considered by adolescents in HCDM	15 youth 12-17 years n=8 JID, n=7 CD)	TNF-xi	Adolescents considered the effects of the treatment to include worsening of symptoms and hope of avoiding side- effects to include quality of life rather than long-term outcomes.
McGrady, M.E., G.A. Brown, and A.L. Pai 2016 Cancer	Exploration of mechanisms that influence daily adherence HCDM	12 AYAs 15-31 years (M=19.19, SD=4.86) (n=7, ages 15-19) (n=5, ages 10-39)	Oral chemotherapy (n=7) or oral antibiotic prophylactic therapy (n=5) compliance	Younger AYAs considered personal short- term goals such as daily activities. While not all understood the purpose of their medication, knowledge as a factor in decision- making while some cited their trust of medical professionals as the reason for choosing to take medication. Emotional support was a positive factor in decision- making. Less compliance was noted with medication side effects.
Needle, J.S., et al. 2020	Exploration to develop a better	10 AYAs, 14-23	Factors influencing	Factors affecting

Bone marrow transplant for cancer	understanding of factors that influence AYA HCDM	years (M=19)	decision-making	consideration for family included the balance of AYA versus family desires. Practical burdens to family such as cost, care work burden was considered. Also, accommodation during treatment, and identification of caregivers. Meaningful time was defined withing quality of life and was considered over hospitalization due to disease.
Pyke-Grimm, K.A., et al. 2022 Cancer	Exploration of involvement of AYAs in daily HCDM	16 AYAs, 15-20 years (M=17.3)	Involvement in day-to-day decision making	Mental mindset describes AYA diagnosis acceptance and its meaning resulting in a will to fight the disease. They took responsibility for making decisions about their daily care and following through with such decisions. AYAs advocated for their health management

				needs through compliance and behavior. There was a choice made regarding others becoming involved in their decision-making, often including parents.
Silwinski, S.K., et al. 2017. Familial hypercholesterolemia	Exportation of challenge related to transition to adult care when assuming responsibility for self-management	12 young adults 17-21 years (M=18.4 ±2.2) and parent dyads	Cholesterol screening and treatment for disease	Most young adults expressed a desire to become primary decision makers but felt a need for parent input due to lack of disease knowledge
Weaver, M.S., et al. 2015 Cancer	Exploration of adolescent preferences ,and parent and clinician behaviors supporting HCDM	40 adolescents 12-18 years	Adolescent involvement in cancer decision making	65% preferred an active role in decision making that ranged from shared decision making to primary decision-maker. Discussions with parents prior to final decision making with the medical team were seen as supportive

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### Chapter III

#### Specific Aims

Sickle cell disease (SCD) affects approximately 100,000 individuals in the United States (U.S.) (Brousseau, Panepinto, et al., 2010). Approximately 1 of every 365 Black or African American infants and 1 out of every 16,300 Hispanic American infants are born each year in the U.S. with SCD (CDC, 2022). Sickle cell disease is an inherited genetic disorder that is marked by recurrent pain episodes as well as organ damage caused by vaso-occlusive episodes (VOE) (Ware et al., 2017). Four phases of pain have been identified in a VOE: two are important in initiation of treatment. The prodromal phase is marked by aches and paresthesia and is present prior to onset of VOE pain. It is during this time, before the initial or the second phase, which shows a slow turnaround of the first phase symptoms. Pain is established and becomes severe, reaching a higher limit. It is at this phase that treatment is recommended (Ballas, Gupta, & Adams-Graves, 2012). The third or established phase lasts approximately three to seven days and is often marked by hospital admission. The fourth or resolving phase lasts eight to ten days. VOE treatment begins at home during the prodromal phase with increased water consumption, heat, over-the-counter analgesics, and opioids (Brandow et al., 2020). VOEs that do not respond to home care require emergency medical treatment with intravenous fluids, oxygen, and intravenous opioids.

Parents/legal guardians are legally responsible for making medical decisions for adolescents under the age of 18 years. Despite recurrent VOEs in this population, it is unknown what level of adolescent involvement is included in the healthcare decision making (HCDM) processes surrounding SCD. An adolescent, defined as being between 14 and <18 years, is not generally considered capable of making medical decisions on his/her

own (Hartman, 2002). However, once the adolescent is an adult, he/she is responsible for making medical decisions. Autonomy-seeking in adolescence is an activity that impacts independence (Spear & Kulbok, 2004). Autonomous HCDM should evolve to become an independent activity as the adolescent/young adult (AYA) transitions to adult health care. This is especially important with chronic illnesses, as with SCD, which may include frequent occurrences of complications and the need to understand the impact on the body and treatment needs. While it is not known how or whether an AYA may be involved in HCDM, it is known that not all parents allow the adolescent to participate in the HCDM process, despite the adolescent being developmentally ready to participate (Miller & Drotar, 2006).

Decisions take into account available options resulting in selection of an option believed to provide the desired outcome (Byrnes, 2002). Collaborative HCDM involves the parent and AYA taking into account activities such as AYA advice-seeking and parent-adolescent negotiation (Miller, 2009). The process of communication in providing needed information (Shersher, Haines, Sturgiss, Weller, & Williams, 2021) and then ascertaining learning is called the interactive communication loop (Schillinger et al., 2003). It is unknown how parents teach, or if they do teach AYAs to make decisions regarding health care and seeking medical care for VOE pain. Adolescent and young adults tend to seek adult input less often as they age, relying on past experience to assist in HCDM (Byrnes, 2002). Autonomous HCDM should evolve during adolescence, becoming an independent activity prior to transition to adult care between the ages of 18 and 21 years, dependent on clinic policy. It is further noted that one must be able to constructively weigh options such as comparison of treatments and understanding of risks, benefits, and potential outcomes.

Autonomous HCDM is important, and adolescents should prepare for this throughout their transition period. The purpose of this study is to examine the thought processes of HCDM by parents/legal guardians and AYAs age 14 to 21 years when considering seeking emergency department care for VOE and perceptions of the readiness of the AYA to assume HCDM by AYAs and parents/legal guardians.

**Aim 1. Identify and compare factors that influence the parent/legal guardian and AYA's decision making when deciding to go to the emergency department for VOE in SCD.** Data will be collected using semi-structured qualitative interviews guided by self-dialogue theory.

**Aim 2. Compare and contrast parent/legal guardian and AYA perceptions about the adolescent's AYA's ability to appropriately seek emergency department care for VOE in SCD.** AYAs and parents/legal guardians will participate in separate semi-structured interviews about the decision to seek emergency department care for VOE.

## **Background**

### **Sickle Cell Disease and Pain**

Sickle cell disease (SCD) is a genetic disease that replaces glutamic acid with valine, resulting in a sickled hemoglobin (HbS) (Bunn, 1997; Lonergan, Cline, & Abbondanzo, 2001; Piel et al., 2017). Homozygous SCD results from the inheritance of two copies of HbS. Double heterozygous SCD is the result of HbS pairing most often with a C, D, E, or Beta thalassemia gene to form other genotypes (Piel et al., 2017). Sickle cell disease affects approximately 100,000 individuals in the United States (Brousseau, Panepinto, et al., 2010; CDC, 2022; Hassell, Eckman, & Lane, 1994). Approximately 1 out of every 365 Black or African American infants and 1 out of every 16,300 Hispanic

American infants are born each year in the U.S. with SCD (CDC, 2022). Sickle cell disease is most prevalent in those of African, Mediterranean, Indian, and Middle Eastern descent worldwide (Ellison & Shaw, 2007; Lonergan et al., 2001).

Sickle cell disease is marked by recurrent pain episode and organ damage caused by vaso-occlusive episode (VOE) (Piel et al., 2017). Vaso-occlusive episodes occur when red blood cells become sickle shaped due to deoxygenation resulting in polymerization of the red blood cell (Edwards et al., 2005; Novelli & Gladwin, 2016). These sickle-shaped cells block small vessels and result in infarction and ischemic damage to surrounding tissue that causes VOE pain in the affected area (Bunn, 1997; Lonergan et al., 2001; Piel et al., 2017). Pain is the most frequent symptom associated with morbidity in SCD (Platt et al., 1991) and is most frequent in patients with HbSS and Hb $\beta^0$  (Stuart & Nagel, 2004) Patients most often complain of back or long bone pain (Field, Knight-Perry, & DeBaun, 2009). Increasing age of children and adolescents correlates with increased VOE episodes (Darbari et al., 2012).

Four phases have been identified in a VOE; two are important in initiation of VOE treatment. The prodromal phase is marked by aches and paresthesia and is present prior to onset of VOE. It is during this time, before the initial phase, that treatment is recommended though it is not frequently initiated at this point (Ballas et al., 2012). The initial phase lasts one to three days and begins at the onset of the VOE. The second phase lasts approximately three to seven days and is often marked by hospital admission. The fourth phase, resolving phase, lasts about eight to ten days. Vaso-occlusive episode treatment begins at home with non-pharmacological care, including water consumption, heat, distraction, and rest (Brandow et al., 2020). Over-the-counter acetaminophen and non-steroidal anti-

inflammatory medications then opioids are administered as needed. A study exploring home care related to pain management reported use of analgesics 85% of days children and adolescents experienced SCD pain (Dampier, Ely, Brodecki, & O'Neal, 2002). When exploring SCD related pain in children and AYAs, ages 6-19 years, an increase in pain severity that resulted in a corresponding increased use of analgesics as children became older was demonstrated (Dampier et al., 2002).

Not all VOEs respond to home care. Those that do not respond to home care require emergency medical treatment with intravenous fluids, oxygen, and intravenous or intranasal opioids (Stinson & Naser, 2003). VOEs can last for several days or weeks (Ballas et al., 2012). Most hospitalizations related to SCD are due to pain from VOE (Cacciotti, Vaiselbuh, & Romanos-Sirakis, 2017). In a study of health care utilization by Brousseau, Owens, et al. (2010), which included 3605 patients ages 10 to 17 years, there were 0.68 (CI 95%, 0.63-0.73) emergency department encounters per patient per year. There are no diagnostics available to measure the existence or severity of VOE. The healthcare provider must therefore portray a sense of understanding and trust in the patient who is reporting pain and offer treatment accordingly (Field et al., 2009).

### **Decision Making**

Decision-making capacity is defined by the processing of information and comprehension by the AYA with an understanding of the illness and proposed treatment information provided by the healthcare provider (Ruhe, Wangmo, Badarau, Elger, & Niggli, 2015). Additionally, decision-making capacity considers the difficulty of the decision and requires the adolescent to clearly communicate his/her decision. Capacity

varies between small choices and more important decisions and may vary depending on experience with HCDM and maturation level.

### **Decision-making by Adolescents**

Adolescents and young adults (ages 14-<21) experiencing a variety of chronic illnesses are required to make daily decisions in the management of their diseases. These decisions may not always be in collaboration with the parent/legal guardian. Type I diabetes is an example of such a chronic disease (Miller & Drotar, 2006). Adolescents must make decisions about nutrition, activities, and indication of possible symptoms of hypo- or hyperglycemia. Such decisions are not always discussed with the parent/legal guardian. Adolescents with Type I diabetes require disease knowledge and HCDM skills to act on daily activities. Adolescents, in a study of mothers (n=84, age range 30.2-60.2, M=48.6[SD5.7], 97% Caucasian) and their adolescent (age range 11.5-17.5, M=14.16[SD1.7], 53% male), collaborating with their mothers regarding type I diabetic care was explored (Berg, Schindler, & Maharajh, 2008). Adolescents, as age increased, tended to seek out collaboration less often, believing they were cognitively more capable of making decisions. Mothers were found to agree that less collaboration was necessary to compensate for cognition as adolescents aged. The best way for an adolescent to learn self-care and HCDM is through collaboration with a parent/legal guardian, who should start at a younger age.

Parent-adolescent communication in daily decision-making regarding treatment of Type I diabetes was studied employing 62 parent-adolescent dyads (Miller & Drotar, 2006). Age of adolescents was 11-17 years (M13.3, SD1.77), 54% were male, and 87% were Caucasian. Parents indicated that higher negative communication by the adolescent

resulted in lower adolescent adherence to care ( $r = -.28, p < .03$ ), and providers reported lower adherence by adolescents ( $r = -.26, p < .04$ ). Further findings indicated that adolescent-parent communication regarding diabetes management resulted in hypervigilance or buck-passing by the adolescent, indicating that discussion of self-care produces negative results. These findings indicate that more research is needed related to parent-adolescent communication regarding chronic illness to ascertain the best way for parents to interact with their adolescents. It also indicates that parents may need to start conversations about self-care prior to adolescence to prevent some negative care discussions and increase adolescent autonomy. While adolescents want autonomy in diabetic HCDM, they may want to work in collaboration with parents when there are other medical issues present.

Adolescents want to participate in HCDM in conjunction with the parent and physician as related to biological therapies (Lipstein et al., 2013). In a qualitative study exploring desire to participate in medical decision making, seven adolescents with Crohn's disease and eight with idiopathic arthritis, nine females, and age range 12-17 years were interviewed. Desire to be involved in HCDM was especially true if the decision pertained to quality-of-life outcomes. In a study examining AYA self-efficacy, decision making, and readiness for transition to adult care, Varty et al., 2022, found that AYAs with high scores in the Decision-Making Involvement Scale related to behaviors of information gathering and advice seeking expressed less responsibility for self-care and self-efficacy. Adolescents and young adults who scored higher on behavior of expressing thoughts and providing information to parents, scoring higher on the Decision-Making Involvement Scale wanted lower parent involvement in self-care and self-efficacy. It is important to understand AYA

decision-making to elicit their desire for self-care and self-efficacy in transition to adult care preparedness.

The inclusion of the AYA by parents in HCDM varies based on medical care needs and expected treatment outcomes. Some parents excluded or believed that adolescents should be excluded from HCDM in some chronic or life threatening diseases such as cystic fibrosis and cancer (Angst & Deatruck, 1996; Varma, Jenkins, & Wendler, 2008). Others contemplated child development as a consideration for including the adolescent in the HCDM process (Geller, Tambor, Bernhardt, Fraser, & Wissow, 2003; Wangmo et al., 2017) or type of decision to be made (McRee, Reiter, & Brewer, 2010).

A qualitative study interviewed 15 parent/legal guardian and adolescent dyads regarding decisions about administration of tumor necrosis factor- $\alpha$  inhibitors treatment for juvenile idiopathic arthritis or Crohn's disease (Lipstein et al., 2016). Side effects of treatment as well as the potential for worsening symptoms and resultant quality of life were considered by parents/legal guardians and adolescents though different factors were considered, with parents/legal guardians reporting concern over long-term effects while adolescents were more concerned with immediate effects. Results of this study indicate that understanding of information is not equal between the parent/legal guardian and adolescent. More exploration is needed regarding factors of importance to adolescents versus parents/legal guardians as well as the context in which they are considered.

While shared decision-making collaboration may result in joint treatment decisions, it may also result in improvement of health. Mental health issues may be acute or chronic in children and adolescents. A study involving 177 children ages 6-12 years (31%) and adolescents ages 14-18 years (69%) examined the impact of shared decision making on

child and adolescent health outcomes (Edbrooke-Childs et al., 2016). Many issues are related to child health (88%) with 12% related to parental needs. Questionnaires were collected from parents, children/adolescents, and/or clinicians at two time points. The purpose of this study was to examine shared decision making of parent and child/adolescent resulting in child/adolescent function. While clinicians did not note any improvement in health, the parents and children/adolescents noted a correlation between shared decision making and clinical improvement of the child or adolescent. Collaboration had a positive perception of health outcomes by children/adolescents, indicating that a collaborative relationship when discussing care and making decisions changed perception of health in a positive direction. Further research is needed to explore ways in which shared decision making affect perceptions of care and health outcomes. Children and AYAs with various chronic diseases have expressed varying degrees of interest in HCDM participation (Coyne et al., 2014; Coyne & Gallagher, 2011; Dunsmore & Quine, 1996; Ellis & Leventhal, 1993; Garth et al., 2009; Geller et al., 2003; Knopf, Hornung, Slap, DeVellis, & Britto, 2008; Mitchell, 2014; Vaknin & Zisk-Rony, 2011). Most who wanted involvement in HCDM expressed a need to have physician and parent input (Garth et al., 2009; Jedeloo et al., 2010; Knopf et al., 2008). This study also noted that a few wanted to make the final decision on their own related to adolescent decision-making skills and autonomy (Ellis & Leventhal, 1993; Geller et al., 2003; Jedeloo et al., 2010). Perception of inclusion was frequently related to type of decision and directly correlated to disease severity and trajectory and often centered on minor decisions such as diet and timing of activities (Angst & Deatrick, 1996; Runeson, Elander, Hermeren, & Kristensson-Hallstrom, 2000; Stegenga & Ward-Smith, 2008; Vaknin & Zisk-Rony, 2011). Those with a life-threatening illness or a need

for surgery frequently felt a limitation to their ability to say no to treatment ((Angst & Deatrick, 1996; Deatrick, 1984; Ellis & Leventhal, 1993).

In addition to cognitive and affective development, adolescents develop health behaviors that will positively or negatively impact future health (Holmbeck, 2002). Parent-child relationships may also impact health behaviors, specifically adherence, based on development of autonomy (Holmbeck, 2002). Chronic diseases may have a negative impact on adolescent development, resulting in a delay of said development (Holmbeck, 2002). Those with SCD may also have impaired cognitive development due to overt stroke, silent stroke, or other reasons related to SCD (Schatz et al., 2002). There have been no studies on the impact of cognition on adolescents or adults with SCD related to decision-making capabilities.

Collaborative HCDM for treatment of VOE pain in the home setting involves the parent/legal guardian and AYA, taking into account activities such as AYA advice-seeking and parent- AYA negotiation (Miller, 2009). Adolescents and young adults tend to seek adult input less often as they age and rely on past experience or peers to assist in HCDM (Byrnes, 2002). Not all parents allow the adolescent to collaborate in the HCDM process, despite the adolescent being developmentally ready to participate (Miller & Drotar, 2006). Knowledge should be translated into usable behaviors that have been learned by the parent and are being taught to the adolescent based on cognitive ability. Knowledge from research and healthcare provider teaching can be translated into workable behaviors (Straus et al., 2011), thus the healthcare provider should have taught the parent from the time of diagnosis how to care for and make decisions about healthcare for the child.

Different experiences and perceptions are based on the role of participant in the phenomena being studied (Patton, 2002). Thus, a primary caregiver and an AYA may perceive the collaborative process of HCDM in different ways, describing reality and experiences from two very subjective perspectives. Both perspectives are real but do not indicate that one is more true or important than the other. Each participant has constructed their reality based on personal experiences and implications the phenomena may have on their individual lives.

### **Dialogical Self Theory**

Dialogical Self Theory explains the self in relation to the thought process with HCDM. The self is composed of two parts: one part of the self is located within the mind and the other part extends to the environment (Hermans, 2012). This can be thought of as what is 'me' and what is 'mine'. The 'me' in this description is a part of the self while the 'mine' is that which belongs to me. Such thoughts came about because of the work of William James, who developed self-dialogue through pragmatic methods (James, 2019).

Dialogical Self Theory (Wray-Lake et al., 2010) will be used in this study to examine internal conversations related to the decision-making process. Exploration of this process related to visiting the emergency department for a VOE due to SCD may aid in understanding how AYAs process thoughts related to HCDM and indicate needed teaching prior to transition to adult care. A decision takes into account available treatment options and behaviors related to selection of the option believed to provide the desired outcome (Byrnes, 2002). Decisions regarding home care for VOE, including non-and various pharmacological treatments and various pharmacologicals, are required from the onset of the VOE. It is unknown how these decisions for home care or presentation to emergency

department or day hospital are made and who makes them: the parent/legal guardian, AYA, or both.

Mikhail Bakhtin added the concept of the involvement of multiple voices to inner dialogue as cited in Hermans (2012). These multiple voices are portrayed as thoughts within the mind of the person making the decision and may potentially include parents/legal guardians, physicians, and community. Collective voices or thoughts are influenced by professionals, jargon, culture, social circles, teachings, and other such entities. As a result of this community of voices, the decision maker may have conflicting thoughts about decisions to go to the emergency department. The culture of medicine and the voice of the healthcare provider may influence the self to seek care. Thoughts about what the healthcare provider taught and how the AYA was treated in the hospital may work to dominate the discussion with the parent/legal guardian. These voices, both as part of the self and as belonging to me, become a story, agreeing, and disagreeing, and developing a solution to the discussion at hand based on possessed knowledge and experience.

### *Part of Self or “me”*

#### **Adolescent and Young Adult**

The ‘me’ can be described as an AYA who has a culture and personal experience (Figure 3). The AYA may be developing HCDM skills, cognitively and emotionally progressing, and is developing autonomy. Adolescents and young adults develop the ability to reason and make decisions as they progress from childhood to adulthood (Dahl, 2004). Piaget noted that the cognitive process, which is used in reasoning and rationalizing when making healthcare decisions, develops over time as infants transition to childhood and then to adolescence (Huitt & Hummel, 2003). Cognitive development involves problem

solving, increasing capacity to make decisions, and development of thought beginning in childhood through to adulthood. It is possible for children of approximately 7-11 years to have experiences in which they apply a logical thought process to actual problems, noted as Piaget's stage of concrete operations (Straus et al., 2011). The next stage is that of formal operations and is applied to adolescents of the approximate ages 11-15 years (Wadsworth, 1996). As cognition develops through this stage, adolescents become capable of imposing logical reasoning when evaluating medical decisions for self-care. Social knowledge, such as culture, in cognitive development involves interaction with others (Wadsworth, 1996). Such knowledge would be used in HCDM collaboration with parents/legal guardians. As adolescent continue to mature, they develop a formal operational stage, which is used by many high school graduates in industrialized countries (35%) (Huitt & Hummel, 2003). Formal thought involves what is real in the world as well as that which is possible (Kazi & Galanaki, 2019). Formal thought allows for consideration of multiple factors and perspectives, such as from parents/legal guardians, experiences, and physicians.

The self is a symphony of inner beings, each with independent viewpoints and cultural guidance. The self does not function independently but functions, instead, as a society. As with any society, the viewpoints of the self may diverge or conflict as the decision being made is considered. This internal dialogue is often less formal and somewhat muted in comparison to external dialogue with others (Hermans, 2012; Hermans, Kempen, & Van Loon, 1992). While external dialogue involves discussion between two or more individuals, inner dialogue entertains a much more complex system of interaction. Included in such internal discussion are memories and experiences such as smells, tastes, sounds, imagery, and tactile stimulation (Hermans, 2012). Such memories

and experiences for the AYA with SCD may be responses of the medical staff to his/her complaints of pain, the discomfort of an intravenous catheter stick, or the feelings that arise from the intravenous narcotic. Based on strength of the voice during inner dialogue, some voices may be suppressed, resulting in a dominant voice that directs the dialogue and decision. This may occur internally when the AYA has bad memories of a previous emergency department visit.

Individuals make decisions based on cognitive ability and their judgment regarding a decision. It should be noted that issues such as impulsivity in relation to psychosocial maturity may impact their decision (Steinberg & Cauffman, 1996). Applying such factors when deciding to seek emergency medical treatment, AYAs would implement autonomy and self-reliance in noting their ability to make the decision while thoroughly evaluating the situation. They would also consider the larger picture, which in the case of VOE pain, would be the care rendered beginning at onset of the VOE and the potential for continued homecare to relieve the pain versus transfer to emergency department care.

Decision-making behavior may be influenced by emotion. Cognitive and emotional development occur at different rates in adolescents (Steinberg, 2005). Piaget also indicates that intellectual development involves an affective component (Wadsworth, 1996). Affective development involves that of emotions and values, which are involved in the decision-making process. Cognitive behavior is influenced by affect (Wadsworth, 1996).

Emotions can positively or negatively affect decision-making, a process that is influenced by similar past experiences (Schwarz, 2000). Thus, results of prior similar decisions regarding emergency medical care for VOE pain influence the current decision based on feelings related to positive or negative outcomes of the emergency medical care

experience. There are two types of emotions that impact the decision, expected emotions and immediate emotions (Loewenstein & Lerner, 2003). Expected emotions are those that are expected because of the decision, such as a reprieve from the pain and having it addressed in a positive manner in an emergency care setting. Immediate emotions involve the feelings that emerge when the decision is made, such as relief that the AYA no longer needs to sit at home in pain.

Emotions may be further affected by two types of empathy gaps, “hot-to-cold” and “cold-to-hot” experiences (Loewenstein, 2005). In the “hot-to-cold” state, individuals are readily influenced by the situation and are not aware of the impact of their emotions on the decision. This may occur in those suffering VOE pain when they consider a bad experience in the emergency department, choosing not to repeat that experience by avoiding the emergency department and treating their pain at home. They are not totally aware that emotions are influencing their decision. On the other hand, those in “cold-to-hot” states may make future decisions based on rationalization, not considering past good or bad experiences.

### **Culture**

The culture of the black community may bring stigma of being black or having SCD while being treated in a medical facility. Stigma often relates to race, ethnicity and socioeconomic status, leaving the individual feeling disapproval or disqualified from acceptance in society (Weiss, Ramakrishna, & Somma, 2006). Health related stigma results from judgment or lack of social acceptance and may result in delayed care-seeking and lead to lack of continuity of care or refusal to continue care (Weiss et al., 2006). This

stigma can come from healthcare providers and others with whom one is associated while seeking care at a medical institute (Bulgin, Tanabe, & Jenerette, 2018).

Stigma with SCD is also seen with reduced access to comprehensive care and is seen more often with SCD than with other genetic diseases (Kauf, Coates, Huazhi, Mody-Patel, & Hartzema, 2009). Sickle cell disease patients have often been judged as being a drug seeker or addict, resulting in healthcare providers discrediting their pain and providing inadequate care (Bulgin et al., 2018). The diagnosis of SCD is also stigmatizing and many families do not share the diagnosis with family and community.

Family, community, and cultural influences as well as access to education impact the health of AYAs (Viner et al., 2012). Adolescents and young adults with SCD may not have access to a support group within the community. Schools spend a limited amount of time teaching about various diseases and health concerns. There may not be much teaching regarding SCD. Extended family may not understand SCD and thus do not know how to be supportive of an invisible disease.

The AYA must also deal with the social determinants of health. Availability of resources, such as good nutrition and housing which may not be close to a health care facility, are affected by income. Level of education correlates with knowledge and decision-making regarding health behaviors. Race and ethnicity may also negatively impact health, as many black individuals and Latinos are more likely to live in poverty-stricken neighborhoods (Braveman, Egerter, & Williams, 2011). While the voices involved in inner dialogue may emerge within thought as individual viewpoints, they may also merge and combine various issues, such as culture, to come to one consensus (Hermans, 2012). When deciding, the AYA may tap into any or all voices. while considering options for

VOE care. Communication with physicians regarding health status and HCDM desires should take into account several factors related to demographics, such as access to transportation, (Makoul, 2001), frequently referred to as social determinants of health. Such determinants include socioeconomic status, education, and race. Health is negatively impacted by below high school graduate education and poverty income levels (Braveman, Cubbin, Egerter, Williams, & Pamuk, 2010).

Other issues related to low income include residential location. Not all families can afford to live in areas that are conducive to lessening sickle cell pain. Research has demonstrated that low humidity and cold weather negatively impact the frequency of sickle cell pain (Jones et al., 2005). Additionally, low income housing is often located in areas with limited access to adequate health care, which may limit ability to seek out emergency medical care (Tucker, 2002). Despite this, due to disease severity, low-income children with SCD were found to significantly ( $p < 0.0001$ ) seek out more emergency inpatient, and home health care than other children with a similar income level (Raphael et al., 2009).

There is a paucity of research related to race and social issues within the sickle cell population (Bediako & Moffitt, 2011). Education in school as well as media coverage of SCD are the most frequent learning methods, both of which are limited and often portray SCD as an African American disease (Wailoo, 2014). Association of cultural beliefs and biases may influence perception of those with SCD as being an African American disease. This may, in turn, affect medical care and public health efforts to address the needs of this population (Bediako & Moffitt, 2011).

Cultural sensitivity to the needs of AYA s with SCD is essential when providing quality health care (Schwartz, Radcliffe, & Barakat, 2007). A culturally sensitive cognitive-

behavioral pain intervention using four sessions to build collaboration between support persons and the AYA with SCD as well as training of the AYA to cope with pain episodes was found to have a positive effect on AYA pain management skills.

### **Experiences**

By the time a child enters the stage of AYAs, they most likely have experienced many episodes of pain which are often addressed in the emergency department. In a study by Kanter et al. (2020) studying adolescents and adults ages 15-50 years, it was found that participants perceived emergency department care to be good quality in only 50.9% (146 of 287) of visits. Participants also noted that they were not adequately prepared during the transition phase to adult care, and did not perceive they could care for themselves appropriately as adults. Kanter et al. (2020) further evaluated frequency and experience of adolescents during the 12 months prior to the study. Sixty-six percent of participants visited the emergency department with 39% reporting neutral experiences and 29% reporting negative experiences. Findings also included a report that 52% of participants reported past bad experiences in the emergency department influenced their decision to avoid emergency department care. Past negative experiences in the emergency department resulted in a reported 67% delay in seeking medical care or avoidance of the emergency department during the prior 12 months.

### ***Belongs to Me or ‘mine’***

The voices within the ‘mine’ of the self-arise from those things that are mine, such as my parent/legal guardian, my doctor, my health, and my peers (Hermans, 2012). External dialogue may involve one or more of those that are ‘mine’. During the event of HCDDM related to visiting the emergency department for VOE pain, the conversation will

take place between the parent/legal guardian and the AYA. Other voices from past discussions, e.g., healthcare providers, may present within the internal dialogue of self and impact the AYA decision. The voices have varying influences on the decision at hand. Some voices may be suppressed within self-dialogue due to having a weakened connection as it relates to influences from others, e.g., my healthcare provider. The voices of others from their social and medical circles may dominate and be powerful due to said influences, taking over the dialogue and the decision. When one voice does not overtake the decision, there may be internal conflict.

### **My Body**

Health, pain management, and other forms of self-efficacy may be associate with that which 'belongs to me' or "mine." Crosby, Joffe, Peugh, Ware, and Britto (2017) studied self-efficacy in 22 AYAs ages 16-24 years related to SCD. In addition to measuring self-efficacy, they measured health related quality of life. After using the Stanford Chronic Disease Self-Management Program there was no improvement noted immediately after the intervention and there was no significant improvement at three months post intervention regarding perceived self-management behaviors. Goldstein-Leever, Peugh, Quinn, and Crosby (2020) also measured self-efficacy and health related quality of life in 101 AYA participants. They found that disease self-efficacy and disease severity were both statistically significant predictors of health-related quality of life with increased self-efficacy in SCD. The issue with studies measuring self-efficacy related to self-management is that decision-making regarding such skills is not measured. While AYA s may perceive themselves as being efficacious in self-management, the HCDM process is not factored into the process of accessing such skills.

**My Parent/legal guardian**

First and foremost, the adolescent may or may not be encouraged to participate in the HCDM process by the parent/legal guardian. Per the legal system, the adolescent, defined as an individual between the ages of 14 and <18 years, is considered generally not capable of making medical decisions on his/her own (Hartman, 2002) but makes no reference to assent. Adolescents are not permitted to give legal consent regarding chronic disease care regardless of how often they experience events requiring similar decisions and must rely on their parent or guardian to do so until the age of 18 years. For these, or other personal reasons, the parent/legal guardian may deny the adolescent's permission to participate in the decision. Exploration of the thought process of the AYA, regardless of whether he/she can participate is important to explore given the AYA will be responsible for making decisions as an adult, even if he/she seeks the advice of a parent/legal guardian.

Within self-dialogue, there is an element of uncertainty which can be complex due to varied positions within the self, ambiguous as a result of delay of understanding, and unformed, or unpredictable (Hermans, 2012). Decreasing uncertainty may be merely a matter of conceding to an external voice from a powerful, and sometimes, dominating others, such as a parent/legal guardian. It can also be reduced by outlining the boundaries between the self and the external individual; something that may not be easily managed if the parent/legal guardian does not grant autonomy. Ironically, internally adding positions may also decrease uncertainty by adding comfort or structure to the previous positions within the self.

It is unknown how confident AYA s with chronic disease feel when making self-decisions or how confident the parent/legal guardian feels in allowing such decisions to be

made. Confidence is “a feeling of having little doubt about yourself and your abilities” (Cambridge, 2019). Age of the AYA, context in which a decision is being made, and cognitive development all factor into AYA competence when making a decision (Grootens-Wiegers, Hein, van den Broek, & de Vries, 2017). Some decisions are also harder than others because of the type of decision to be made or because the adolescent does not feel confident to make the decision (Fischhoff, 2008).

Preferences for method of HCDM among AYAs and parents/legal guardians should be determined in the clinical setting. Knopf et al. (2008) explored parent and adolescent preferences for decision-making when dealing with chronic disease. Adolescents (n=82) with cystic fibrosis (15%), juvenile rheumatoid arthritis (45%), inflammatory bowel disease (27%), and SCD (16%) were recruited. Mean age was 15.4 years with 45% being male, 83% being white, and 17% being black. There were 62 parents recruited. Passive HCDM, as measured using Preferences for Decision-making Involvement, in which the physician makes the medical decision was most often preferred by both adolescents (46%) and parents (53%) while shared decision making was preferred by 37% of adolescents and 36% of parents. Within each group, adolescents and parents, there were significant differences in HCDM style preferences. However, when comparing adolescents as a group to parents as a group, there were no significant differences. Further exploration, however, indicated that only 33% of parent/adolescent pairs agreed. This research indicates that the differences between and within parent/adolescent groups need further exploration into why there are such differences, especially considering the findings that parents and adolescents were largely not in congruence with HCDM preferences. While passive HCDM may be possible in the clinical setting, parent/legal guardian and adolescent collaboration in the

form of shared decision making in the home setting when managing chronic illness is required.

### **My Doctor**

Past decisions with parents/legal guardians may not be the only ‘mine’ voices to affect conversation within the self. Despite the healthcare provider not being present during the discussion between the parent/legal guardian and AYA at home, the inner voice of the healthcare provider may weigh on the self-dialogue process. Past healthcare provider education and discussions regarding self-care during VOE may play in the mind of AYA and influence HCDM. Memories of peer responses may also lend their voice to internal self-dialogue. Comments regarding the amount of school missed or mention of the AYA’s health may impact the AYA desire to be in school versus at the hospital.

### **Conclusions**

Parents/legal guardians and AYAs may not consider similar factors when making decisions about AYA health care. Both parents/legal guardians and AYAs may consider many factors related to treatment decisions and it is not known how each party perceives the concerns of the other. Collaboration in decision-making requires the parent/legal guardian and AYA to work together to make treatment decisions but can prove to be difficult when they are not considering the same factors, such as short- or long-term risks or benefits and personal or cultural beliefs and values. Research has not explored decision-making activities when parents and AYAs are making health care decisions regarding at-home treatment versus emergency department care for VOE in SCD.

Ethically, it is the right of individuals to be involved in medical decision-making as a right of self-determination (McCabe, 1996). Autonomy-seeking in AYAs is a normal

developmental process that impacts independence (Spear & Kulbok, 2004). Autonomous HCDM should evolve during adolescence to become an independent activity as the AYA transitions to adult health care. This is especially important with chronic diseases, such as SCD, in which there may be frequent occurrences of complications with a need to understand the impact on the body and resultant decision-making needs.

Development of adolescent judgment and decision-making skills are considered to be incomplete and not comparable to those of adults (Steinberg & Cauffman, 1996). Healthcare providers should involve AYAs, beginning at the onset of adolescence, in the process of decision-making to allow them to understand methods of weighing options as well as considering risks and benefits. This will help prepare the AYA for the process of decision-making at home in collaboration with their parents/legal guardians. This will also allow the AYA to determine the preferred style of HCDM he/she desires.

Adolescent and young adult development indicates they are generally capable of participating in decision-making regarding treatment options for health care. AYAs will be required to make decisions as adults after the age of 18 years when experiencing a VOE. It is important when developing positive health behaviors during AYA years that AYAs learn to make good medical decisions prior to transition to adult care. Little is understood about the current ways in which AYAs, and parents/legal guardians collaborate to make health decisions and how such collaboration will impact future decision-making regarding treatment of VOEs in SCD.

## **Methods**

### **Research Design**

This study will use a qualitative descriptive design, which allows for use of theory to guide the study and provides for the ability to understand the context of issues in a given scenario in its natural state (Colorafi & Evans, 2016). Thematic analysis methods will be used to analyze semi-structured interview data of parents/legal guardians and their AYA with SCD. Individual perspectives of two individuals of the same event provide two narratives. Qualitative methods are appropriate because the factors that influence the collaborative decision-making process between parents/legal guardians and AYAs when considering emergency department care for acute VOE are unknown.

### **Population and Sample**

Atlanta, Georgia has one of the largest sickle cell populations in the country with over 3400 child and adult patients (RUSH, 2008). There are three pediatric clinics available in the immediate Atlanta area for AYAs to seek care for SCD. A convenience sample of parent/legal guardian and AYA cohorts will be drawn from patients seeking care for SCD from the Children's Healthcare of Atlanta (Egleston) emergency department. There are approximately 160 AYAs, aged 14 to 21 who regularly obtain care for SCD at this hospital. For this study, saturation will be used as a guide for final sample size for thorough data analysis. We expect a total purposive sample size of up to 30; 15 each AYAs and parent/legal guardian within a cohort. A total of 30 participants should provide an adequate number of participants to reach saturation, for an overall (Creswell & Clark, 2017).

### **Inclusion/Exclusion Criteria**

Adolescent and young adult participants will range in age from 14 to less than 21 years of age. They will have a diagnosis of SCD, and all genotypes will be recruited. The parent/legal guardian of the AYA will be 18 years of age or older. All participants must be

fluent in English. Participants will be limited to those self-identified as African American. No exclusion shall be based on sex. Participants will have presented to the emergency department for treatment for acute VOE using intravenous or nasal pain medications; they may be discharged home from the emergency department or be admitted to inpatient care. The parent/legal guardian who comes to the emergency department with AYA, or if both parents present with AYA, the parent whom the AYA identifies as most involved in the decision-making process will qualify for the study.

### **Recruitment**

Convenience sampling will be used as potential participants present to the emergency department for treatment of VOE. Equal number of women and men (parents/legal guardians) are unlikely to be recruited. Mothers most frequently accompany AYA SCD patients to the emergency department and are their primary caregivers in most instances. Most patients with SCD are of African American descent thus this study will limit participants to those who consider themselves to be African American. It is anticipated that a relatively equal number of male and female AYA s will be recruited given that SCD affects both sexes equally.

Permission to recruit from the hospital within the Children's Healthcare of Atlanta pediatric system will be obtained from the administrative emergency department doctor, Dr. C, Morris. Approval of the proposal and recruitment plan will be approved by the SCD Protocol Review Committee and the AFLAC Protocol Review as well as Children's Healthcare of Atlanta and University of Missouri IRBs. Permission to access admission lists of SCD patients admitted to the emergency department for SCD VOE will be obtained.

Parents/legal guardians and patients will be contacted via telephone call within one week of the emergency department visit to discuss the study.

The investigator will meet with emergency department research staff prior to initiation of the study. A summary of the study will be provided via PowerPoint for those in attendance as well as extra copies for those unable to attend. PowerPoint will include study purpose, study activities, and inclusion/exclusion criteria. Contact information (Emory email) for the investigator will be provided. The emergency department research staff will not be expected to use all inclusion/exclusion criteria to rule patients in or out, just to identify potential patients. If the AYA qualifies based on limited screening by the emergency department research staff, the parent/legal guardian and AYA will be told there is a study about how AYAs make decisions about coming to the emergency department and asking their permission to be contacted by the investigator. A flyer about the study with DR's contact information will be provided for each potential subject. The emergency department research staff will notify the investigator via secure SharePoint of patients who refuse or agree to be contacted. If they agree, the admission date and the parent's/legal guardian's phone number will be provided to DR via SharePoint, and whether they were admitted to inpatient or discharge to home. DR will also contact the emergency department staff up to 4 times per week as needed to ascertain if there were any patients interested in being in the study. Additionally, the SCD inpatient unit will be monitored for admissions for VOE. DR will go to the department and ask the nurse assigned to the patient if DR can approach them about a sickle cell research study. The investigator will then review the electronic medical record using partial HIPAA waiver for full inclusion/exclusion criteria. If the AYA qualifies, the parent/legal guardian will be contacted via telephone by DR. If

the AYA is 18 or older they will be contacted directly by the investigator for potential recruitment to the study. A phone consent script will be read to the AYA 18 or older who was a patient or the parent/legal guardian who attended the emergency department visit with the AYA to determine potential interest in the study. If more than one parent/legal guardian attends the emergency department visit with the AYA, the parent/legal guardian that the AYA identifies as helping most with the decision to visit the emergency department will be asked to enroll. If the parent/legal guardian does not answer the phone, a text and voice mail will be left stating that DR is from Emory/Children's Healthcare of Atlanta and would like to discuss a possible research study involving the study of the parent/legal guardian and AYA. DR will attempt to contact the parent/legal guardian on three separate days. If the parent/legal guardian provides permission, adolescents under the age of 18 will be read the telephone assent. If the adolescent is not home, DR will request a timeframe in which to reach out to the adolescent on his/her phone or a time to call the parent's/legal guardian's phone back to reach the adolescent. If both are interested in the study, the parent/legal guardian and the adult AYA will be verbally consented and the adolescent <18 years will be verbally assented. Immediately after verbal consent and assent are obtained, demographic data will be collected from both the parent/legal guardian and AYA. All interviews will be conducted using zoom or telephone, dependent on participant comfort, will be scheduled at a time convenient to parent/legal guardian and AYA. The parent/legal guardian and AYA interviews will be scheduled at separate times. It will be stressed that interviews are to be private between parent/legal guardian and DR or adolescent and DR to maintain confidentiality. If zoom audio is being used and fails during the interview, an audio recorder will be used via telephone to collect the interview. All recordings will be

stored on One Drive through Emory University. Audio recordings will be destroyed according to Children's Healthcare of Atlanta IRB policies.

### **Data Collection**

#### **Demographics**

Demographic information will be collected after consent and assent. Demographic information from parents/legal guardians and AYA s will include sex as male, female, or other; that they consider themselves African American; age in years; and education level as categories based on years of schooling. Additional information will be collected from parents/legal guardians to include household structure (single or two-parent household). Additional demographic information will be collected from the electronic medical record and include current method of SCD treatment, frequency of VOE in past year that required emergency department visit for intravenous or intranasal pain medications with home discharge or hospitalization, and genotype as it is often related to severity of pain and frequency of pain episode.

#### **Qualitative Interviews**

Qualitative interviews will be conducted via zoom or telephone interview at a time convenient to the participant, within 2 weeks of an emergency department visit to reduce issues related to event recall. Parents/legal guardians will be interviewed separately. Separate interviews help to eliminate influences between the two participants. The participant will be informed of recording for the purpose of transcription, use of identification number rather than name and to provide permission to refuse to answer questions that make the participant uncomfortable. A semi-structured open-ended interview guide will be used to ensure all conversation presented by the interviewee is probed. Probes

were developed based on knowledge of VOE by the interviewer and Dialogical Self Theory (Appendix A and Appendix B). Dialogical Self Theory was used to develop semi-structured interview guides for data collection. There are benefits to collecting data from dyads using individual interview techniques (Eisikovits & Koren, 2010). Individual perspectives, while also employing a dyadic perspective, provide two narratives. Interviews will explore how the decision was made to go to the emergency department during their most recent emergency department visit. The interviews explore the thought process during this decision as well as past experiences making such decisions and elicitation of who made the decision (joint or single decision maker). Individual interviews are expected to last up to 45 minutes and will be audio recorded.

Interviews will be collected by the same individual (DR) at different time points convenient to each participant. This allows for the presentation of individual perspective without concern for the reaction of the partner related to the HCDM process. It will also eliminate concerns of being judged or criticized (Morris, 2001). It allows for confidentiality of data. Caution will also be taken when inserting quotes into the manuscript or with member checking such that the family member does not recognize that they are being discussed (Eisikovits & Koren, 2010).

Data analysis is also impacted by individual interviews. Interviews with the same family unit that are conducted separately provide for the opportunity to examine similar and contrasting versions of the decision to go to the emergency department for treatment of VOE pain (Eisikovits & Koren, 2010). This will be completed using thematic analysis while examining individual interviews separately and then examining them through

comparison of themes. This allows for reconstruction of themes as well as the emergence of distinctive subthemes.

### **Data Management**

Interviews will be recorded and transcribed via Zoom and transcription errors will be corrected by DR. Qualitative data audio and transcripts will be stored on One Drive at Emory University and will be available to DR, and the dissertation committee. Audio recordings will be deleted from the recorder after the recording is transferred to the shared drive and data analysis has been completed. The recorder will be locked in the DR's desk at Emory University until the recordings are erased. Data will be analyzed using MAXQDA qualitative analysis software. This will allow for organization of data from AYA s and primary caregivers. Use of MAXQDA will allow memoing to track the meaning of data and maintain accuracy. MAXQDA program has a memoing feature that can be directly linked to the data being coded. One memo may be linked to multiple segments of data or may be linked individually to a single segment of data.

### **Data Analysis**

Descriptive thematic analysis will be used to analyze data. Thematic analysis lends itself to the development of a systematic framework. This allows for pattern identification. Thematic analysis also allows for both inductive and deductive analysis, providing for examination of participant experiences while deeply exploring their meanings (Braun & Clarke, 2006). Analysis of AYA and then parent/legal guardian interviews will occur initially to address Aim 1: Identify and compare factors that influence the parent's and AYA's decision making when deciding to go to the emergency department for VOE in SCD. These analyses will be followed by analysis of interviews from Aim 1 to address Aim 2:

Compare and contrast parent and AYA perceptions about the adolescent's AYA's ability to appropriately seek emergency department care for VOE in SCD. Thus, analysis will occur across AYA interviews, and across parent/legal guardian interviews.

Analysis will proceed using Braun and Clarke (2006) six steps in thematic data analysis.

1) It is imperative to in-depth coding to become very familiar with the data while verifying accurate transcription of audio recordings, and repeatedly reading the transcripts, noting initial thoughts that are related to meaning and patterns emerging within the data. Data analysis will begin by reading each interview while being attentive to the unit of analysis, the individual AYA or parent/legal guardian within the study. Each transcript will be read and re-read, keeping the Dialogical Self theory in mind. It will provide a guide to compare and finalize inductive codes developed during data analysis.

Data will be coded deductively for Aim 1 using basic fragments or sections of information that can be assessed and analyzed in a meaningful way in relation to HCDM of AYAs and parents/legal guardians as it occurs when considering a visit to the emergency department. Deductive coding will be used to develop a priori categories generated based on Dialogical Self Theory with attention to the primary research questions. Inductive coding will then be used to add understanding of what participants said and important data not captured in the deductive coding (Azungah, 2018). These codes will be saved into MAXQDA, which allows for recording of definition. A code book will be developed for AYAs, and a separate code book will be developed for parents/legal guardians to provide for exploration of thought process within each group regarding HCDM. Inductive coding,

developed from the data, will be conducted using these coded fragments and sections of information for Aim 2.

2) Initial coding of words, phrases, or segments of data will be completed, creating a plethora of codes (Braun & Clarke, 2006). Data analysis is an iterative process that involves movement within the data set, the coded segments of data being analyzed, and the analysis of the data that is being constructed (Braun & Clarke, 2006). Patterns may begin to emerge and will be noted in MAXQDA, using the feature of notes. Identification of words includes coding of words, phrases or sentences surrounding the data of interest so as not to lose context or meaning. Codes are specific ideas used to identify that which is of interest in the data in relation to the phenomenon being studied (Braun & Clarke, 2019)

As codes and patterns begin to emerge, semi-structured interview questions may be modified using an iterative process to better capture data and to probe findings, resulting in more in-depth interviews and better clarification of emerging themes. The interview guide will be edited to delete questions that do not provide data, the remaining questions may be edited, or probes added to elicit more in-depth data, and new questions may be asked based on unexpected codes that may be identified. If new codes emerge as analysis proceeds, previous transcripts will be re-reviewed for the presence of the new codes. This will continue to occur until no new codes emerge in the data and findings are becoming repetitive in nature.

Analysis will begin after the initial 2 to 3 interviews in each group, parents/legal guardians and AYAs, and separate code books will be developed (although we may find common codes across the two). The code book will be modified as needed during the analysis phase and completion of member checking. Member checking will involve

presentation of emerging themes and subthemes from each group. Parents/legal guardians will only hear parent/legal guardian themes; AYA s will only hear data from AYA interviews. Input from participants will be gathered to assess appropriate analysis of data and obtain suggestions for what seems to be missing from the analysis to date. This will guide rephrasing of questions that limit the type of data needed as well as the addition of questions to clarify data using an iterative process. The code book will then be finalized. Interviews that were already coded will be re-coded to incorporate newly identified codes. Data will continue to be analyzed in an iterative manner after every 2 to 3 interviews to determine saturation.

3) Data analysis for exploration for themes will now begin (Braun & Clarke, 2006). Themes are developed through a process of collating codes and using codes as primary themes or subthemes. Codes are assigned to various themes or placed in a miscellaneous theme for review prior to final analysis if they initially do not seem pertinent to the data analysis. Themes represent a concept demonstrating a meaningful pattern or a concept that is frequently displayed in the codes and is demonstrative of a central concept or idea (Braun & Clarke, 2019). Tables and maps will be used to assist in sorting and assigning codes to themes. Use of MAXQDA will allow for creation of maps.

4) Themes will be reviewed and checked against phrases, segments and words coded within the data (Braun & Clarke, 2006). Inductive coding will be used to collect phrases, segments and words from the participant perspective. An inductive or latent approach will be used also , noting that it explores and analyzes meaning given to the phenomena and the data from the participant and the researcher perspective (Braun & Clarke, 2006). Themes will be organized into primary themes, which may entail breaking

down initial themes into smaller themes or merging similar themes into one broader theme. Analysis of data segments for each code will be completed. It will be determined if adherence of the data forms a coherent pattern within each theme through review of coded data excerpts. Deductive and inductive codes will be compared and merged as analysis progresses. Further, consideration of the thematic map will validate meanings within the data and assure that the data has been consistently coded (Braun & Clarke, 2006).

After step 4 and in each set of interviews, AYA and parent/legal guardian, comparison of themes will take place to explore agreement and variability of factors between each group to address Aim 2 (Collaço et al., 2021). Additional themes may be developed, reduced, or expanded based on the experience of the two groups.

5) Analysis is an ongoing process, thus further naming and defining of themes may occur (Braun & Clarke, 2006). Each theme has a purpose in the telling of the story presented in the data and all themes must connect to tell the larger story (Braun & Clarke, 2006; Tucker, 2002). As themes are further defined and sub-themes are clarified, sub-themes may be needed as part of a larger them to define its structure (Braun & Clarke, 2006).

6) Writing of the final report, journal article or dissertation occurs after data have been thoroughly analyzed (Braun & Clarke, 2006). The process involves relating the analysis back to the identified quotes or data segments and the data set as well as relating it to the literature and the purpose of the study. Data extracts will be used to explicate the story being told by the data.

Member checking will occur with participants who have been interviewed. The initial three to four participants in each cohort will be contacted by phone after initial data

collection and analysis of transcripts, provided with initial findings, and asked their opinion regarding capture of meanings and themes within the data as related to participant experiences to validate interpretation of data. It is anticipated that they will be contacted within 6 weeks of the initial interview. These discussions will take place within the context of participant personal experience with decision-making regarding the decision to seek emergency department care for acute VOE. This process will occur after the initial 4-6 interviews are coded and a code book has been developed. Those who were not contacted at this time will be contacted during final analysis for member checking. Member checking or discussing findings with the participants during the data analysis process offers validity to findings and is especially useful when doing solo or individual data analysis (Saldaña, 2016). A final codebook will be developed, noting that new codes may be added based on further data analysis and earlier data may need to be reanalyzed.

Data analysis of two groups discussing the same events may be challenging given differing memories or translation of that event. Mapping will be used, creating a timeline for events related to the process of HCDM (Ummel & Achille, 2016) regarding visit to the emergency department. Mapping will involve a diagram of codes identified in the data with lines of direction indicating connection between categories. As analysis develops and themes emerge, categories will be replaced with themes. Such diagrams will visually demonstrate the connections and direction between themes to visually tell the story emerging from data related to HCDM. To maintain rigor in data analysis, reflective journaling will be employed by the investigator (Ummel & Achille, 2016), noting like and different perspectives of the events of decision-making by the primary caregiver and the AYA.

Sound decisions are required in relation to appropriateness of methodology resulting in integrity of results. Audit trails will be established to include decisions and actions related to the analysis, memoing and all other decisions made. Journaling regarding thought process in study design, data collection, and data analysis will be maintained to provide for an adequate audit trail. Memoing will further define these activities and will be used to clarify thought processes and reaffirm purposes for HCDM activities at various points in the study.

There are several threats to validity (Sandelowski, 1986). “Going native” is a concern when researchers become overly engaged with or immersed in the population being studied. This researcher has been very involved with adults who discuss pain episodes while attending SCD conferences. Care will be taken to avoid bringing biases regarding experiences of patients seeking emergency medical care using memoing in a notebook regarding thoughts related to knowledge of adult experiences in HCDM regarding treatment at an emergency room for acute VOE.

### **Limitations**

There are several potential limitations to this study. One such limitation is the inability to identify and interview AYAs/primary caregivers who decide not to come to the emergency department for VOE treatment to understand why the decision was made not to report to the emergency department. There may be low generalizability given that the findings will be based on a small sample size of African American AYAs and caregivers. Participants may respond in ways they anticipate that the researcher may expect rather than the actual decision-making behavior. Although efforts will be made to ensure each family

member is interviewed separately, because the interviews will occur over the telephone or zoom the investigator may not know if others are present during interviews.

### **Ethical considerations**

#### ***Institutional Review Board (IRB)***

IRB approval will be obtained from Children's Healthcare of Atlanta IRB and from the University of Missouri IRB.

#### **Consent and Assent**

It is imperative that research staff address issues of informed consent and assent prior to and during consent/assent. Vulnerable populations are not able to make decisions independently and autonomously or have an issue that may put them at greater risk for coercion (Shivayogi, 2013). Vulnerability may be the result of developmental issues or undue influence by others, such as from parents/legal guardians, medical community, or prison systems. Additionally, vulnerable populations may include cognitively or physically impaired, military personnel, elderly, ethnic minorities, economically or educationally challenged, pregnant women, fetuses and neonates (HHS.gov, 2018). It is important to protect the vulnerable from coercion through risk management and other forms of protection (Shivayogi, 2013). The Nuremberg Code addresses rights of research participants (Shuster, 1998). Participants have the right to be respected as autonomous with the ability to make independent decisions. It is imperative that the participant receives informed consent, knows what his/her rights are in understanding the consent, and is aware of his/her right to say no or to withdraw from the study. It is also imperative that the participant knows what will be done with the data, who will see it, how confidentiality will be maintained, and is able to give a HIPAA authorization (Shuster, 1998; Smith-Tyler,

2007). All issues will be addressed during the consenting process and will be specifically addressed during discussion.

### *Human Subjects*

#### **Protected Health Information**

Name, address, telephone number, and email address will be recorded to facilitate contact with study subjects. No other personal health information will be collected or recorded. Discussion of the study will include purpose, procedures, risks and benefits, and compensation. All questions will be asked and answered.

#### *Third Party Information*

1. There will be no third-party information collected.

#### *Potential Risks*

There is a minimal risk of breach of confidentiality. A HIPAA waiver will be obtained to allow review of medical records for the purpose of verification of qualification to enroll in the study and to collect data regarding type of SCD and number of emergency department visits and hospitalizations during the past year. Participants have the right to refuse to answer self-reported questions regarding their health.

There is a risk of discomfort with qualitative interviews. The participant is not required to answer any question he/she does not feel comfortable answering. The participant may speak with the primary investigator, his/her physician or a member of the healthcare team as needed.

#### **Potential Benefits**

There are no benefits to participating in this study, but findings may be used in the future to assist AYAs and primary caregivers with collaborative decision-making regarding

treatment-seeking for an acute pain episode. It may also indicate educational needs related to treatment-seeking that may be addressed by healthcare providers.

### **Data and Safety Monitoring Plan**

Research staff directly involved in the study will have access to data collected during this study. Paper documents will be filed in a locked filing cabinet in a secure office setting at Emory University. Electronic data will be maintained on a secure Hematology/Oncology share-drive with limited access behind a secure firewall at Emory University. Participant data will be identified with a unique number. The key connecting the participant to the identification number will be maintained separate from the study data and will be destroyed upon completion of the study, to include dissemination of findings. The identification number key will be accessible only to members of the research team. All audio recordings will be identified on the recording with the unique participant identification number. All transcripts will be identified using the participant identification number. Names of parents/legal guardians, patients, and healthcare providers will be indicated with “XXX” in the transcript. Audio transcriptions will be destroyed after transcription is complete and verified, analyzed and findings are disseminated.

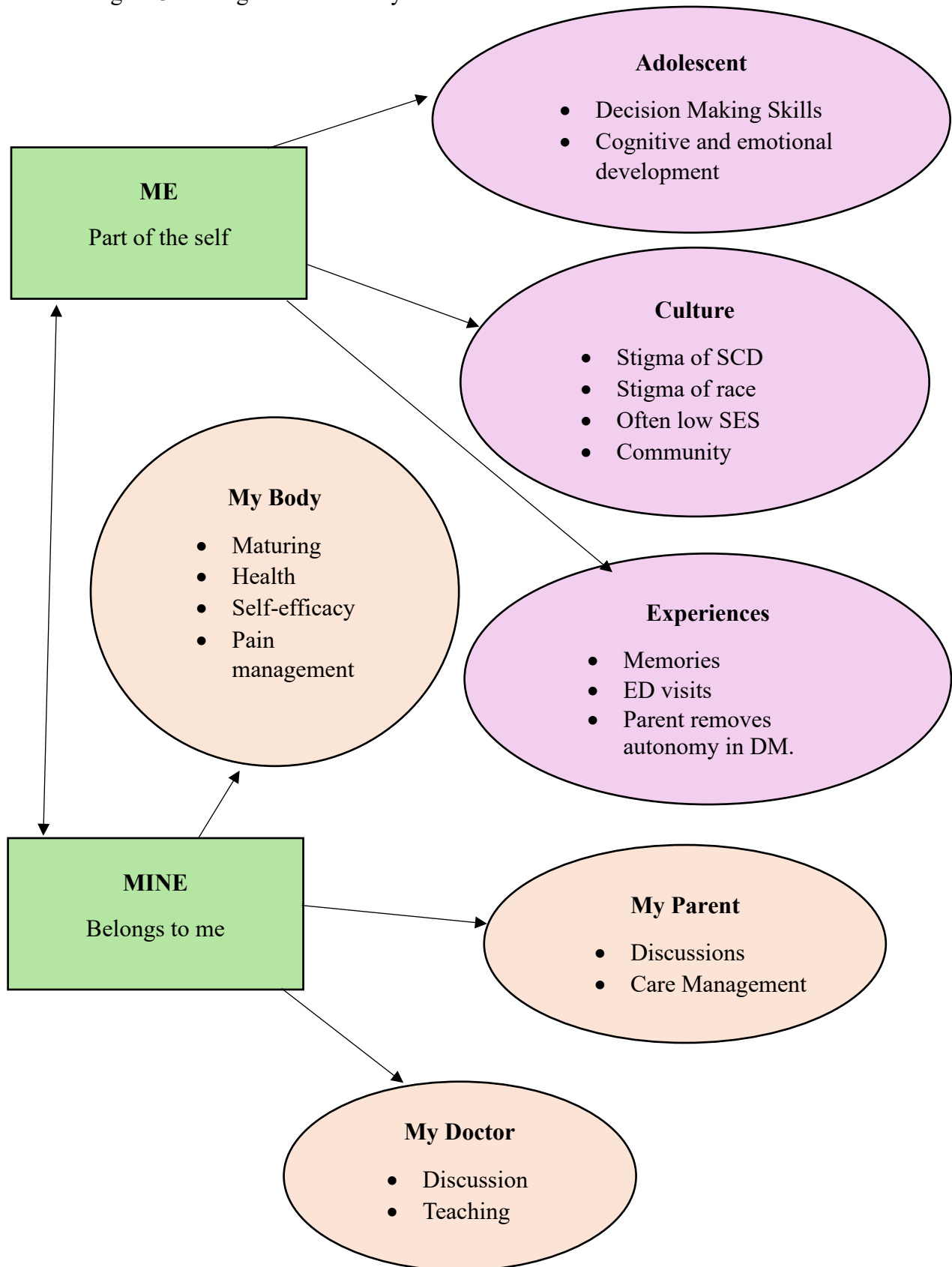
### **Withdrawal from Study**

The participant may withdraw from the study at any time by notifying the PI and/or the research team. All data collected up until the point of withdrawal will be included in data analysis. No further data will be collected. The participant may be withdrawn from the study if the participant does not participate in the qualitative interview.

### **Compensation**

Participants will not be compensated for their participation in this study. There is an audio recorder available for use at Emory University. All costs will be assumed by DR.

Figure 3. Dialogical Self Theory



### Appendix A. Semi-structured Interview Guide Adolescent

So first, I just want to get to know a little about you.

1. What grade are you in this year at school or have you graduated?
  - a. (if graduated) are you in trade school or college or do you work?
    - i. (if so in school) What are you studying?
    - ii. How do you like it?
    - iii. What was your favorite subject in high school?
  - b. (if graduated and work) What do you do for your job?
    - i. What is your favorite part of your job?
    - ii. Tell me about your typical workday.
    - iii. What kinds of things do you do when you are not at work?

Thank you. I want you to think about your most recent visit to the emergency room for a pain episode and treatment. I am going to ask you some questions about how you and your parent(s) decided to go to the emergency room.

2. How do you learn about sickle cell disease?
  - a. Parent teaches
  - b. Nurses, doctors
  - c. Reading
  - d. Internet
  - e. Other
3. Tell me about your last pain episode.
  - a. Triggers
  - b. Describe the pain
  - c. How would you rate the pain on a scale of zero to ten with zero being no pain and 10 being the worse pain you can imagine?
  - d. Did you talk to your parent about how to treat your pain at home?
  - e. Tell me about what you both said
    - i. What were you thinking about when you were talking about treating your pain?
    - ii. Other pain episodes
    - iii. Ways to treat you pain
    - iv. Ways you treated your pain in the past
    - v. Other
    - vi. What did you decide to do about your pain?
      1. Home pain plan
      2. Meds
      3. Water
      4. Heat
      5. Rest

6. Other
  - f. Who made the decisions about how to treat your pain at home?
4. When that treatment did not work, what did you and your parent talk about?
  - a. Tell me about your pain at that time.
5. What were your thoughts when you were talking with your parent about your pain?
6. When did you decide to go to the emergency room?
7. Tell me about your pain at that time.
  - a. Describe
  - b. Rate
8. What were you thinking about when you were deciding to go to the emergency room?
  - a. Tell me what you and your parent talked about when you were making this decision.
  - b. Who made the decision to go to the hospital?
    - i. Tell me what you and your parent talked about when you were deciding
    - ii. What were you thinking about?
      1. Emergency room pain plan
      2. Past emergency room visits
      3. Doctors' advice
      4. What would happen when you went to the emergency room
9. Tell me about your visit to the emergency room
  - a. How long did you have to wait to be seen
  - b. Who described your pain and treatment at home to the medical staff
  - c. What type of treatment did you receive
  - d. How did the staff treat you
10. Tell me about some of the health decisions you make regarding your sickle cell disease?
  - a. Tell me about what you and your parent talked about when you were deciding how to make this decision
11. Tell me who you think should decide how you treat your pain at home
12. Who should decide when you should go to the hospital for your care?
  - a. How old should a child or adolescent be when starting to make decisions to go to the ED for VOC
13. How do you want to learn to make decisions about your care?
  - a. Parent teaches
  - b. Doctors and nurses teach
  - c. Reading
  - d. Other
14. Why do you think you have enough information to make decisions about your sickle cell disease?

15. Do you want to make decisions about your sickle cell disease and why or why not

### **Appendix B. Semi-structured Interview Guide Parent**

So first, I just want to get to know a little about you and your adolescent.

1. Tell me a little about your adolescent or young adult child.
  - a. If in school, what grade are they in?
2. Do you work outside the home?
  - a. What type of work do you do?
3. Tell me a little about a typical day for you.
4. Is your adolescent or young adult still in school?
  - a. Do they work?
  - b. What is a typical day for them?
  - c. Tell me about what happens when your adolescent or young adult has a pain episode.

Thank you. I want you to think about your adolescent's most recent visit to the emergency room for a pain episode and treatment. I am going to ask you some questions about how you and your adolescent decided to go to the emergency room.

5. Tell me what that pain episode was like for your adolescent.
  - a. Probes
    - i. Triggers
    - ii. How does he/she describe the pain?
6. Tell me about their last pain episode.
  - a. How did they describe it?
  - b. Do you know what triggered it?
  - c. What did you do at home for the pain?
    - i. Who decided how to treat the pain?
    - ii. What were you thinking about when you were making this decision?
      1. Ways to treat the pain.
      2. Home pain plan
      3. What your adolescent wanted to do to treat the pain
      4. What worked in the past to treat the pain?
7. Tell me about the things you were thinking of during this conversation to treat the pain.
  - a. How did you go about making that decision?
  - b. Who was involved in making the decision?
    - i. Tell me about the conversation you had with them to make this decision.
8. When the initial treatment did not work, what was the discussion like between you and your adolescent about further treating the pain?
  - a. Opioids, other treatments.

- i. Tell me about your understanding of the pain at that time.
- 9. What were your thoughts when you were having this discussion with your adolescent?
- 10. Tell me about your visit to the emergency room.
  - a. How long did you have to wait to be seen?
  - b. What type of treatment did you receive?
  - c. How did the staff treat you?

Now I want to talk about when your adolescent's pain episode got to the point that you and your adolescent decided that at home treatment was not working.

- 11. Tell me your understanding of your adolescent's pain at that time.
- 12. What were your thoughts when you were considering taking your adolescent to the emergency department?
  - a. Tell me about your discussion with your adolescent when considering going to the emergency room.
  - b. Tell me how the final decision to go to the hospital was made.
    - i. Who made the decision and why?
  - c. What were your thoughts when making this decision?
    - i. Emergency room pain plan
    - ii. Past emergency room visits
    - iii. Doctors' advice
    - iv. What would happen when you went to the emergency room
    - v. Transportation
    - vi. Childcare
- 13. How has and is your adolescent learning about sickle cell disease?
  - a. Parent teaches.
  - b. Doctors and nurses teach.
  - c. Reading
  - d. Other
- 14. Tell me about some of the health decisions your adolescent makes regarding his/her sickle cell disease.
  - a. Who should teach them to make decisions about sickle cell disease and healthcare?
  - b. How should they be taught to make decisions?
- 15. Tell me what you think of his/her decision-making abilities.
  - a. What do you think about how mature they are?
  - b. What do you think is their ability to make decisions about their healthcare?
- 16. How do you believe your adolescent wants to participate in making decisions about his/her sickle cell disease?

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## Chapter IV

Sickle cell disease (SCD) affects approximately 100,000 individuals in the United States (Brousseau, Panepinto, et al., 2010; CDC, 2022). Approximately 1 of every 365 Black or African American infants and 1 out of every 16,300 Hispanic American infants are born each year in the United States with SCD (CDC, 2022). Sickle cell disease is an inherited genetic disorder that is marked by recurrent pain episodes as well as organ damage caused by vaso-occlusion (Ware et al., 2017). Four phases of pain have been identified in a vaso-occlusive episode (VOE): two are important in initiation of treatment. The prodromal phase is marked by aches and paresthesia and is present prior to onset of VOE pain. It is during this time, before the initial or the second phase, which shows a slow turnaround of the first phase symptoms. Pain is established and becomes severe, reaching a higher limit. It is at this phase that treatment is recommended (Ballas et al., 2012). The third or established phase lasts three to seven days and is often marked by hospital admission. The fourth or resolving phase lasts eight to ten days. VOE treatment begins at home during the prodromal phase with increased water consumption, heat, over-the-counter analgesics, and opioids (Brandow et al., 2020). VOEs that do not respond to home care require emergency medical treatment with intravenous fluids, oxygen, and intravenous or nasal opioids (Stinson & Naser, 2003).

Parents/legal guardians are legally responsible for making medical decisions for adolescents under the age of 18 years. An adolescent, defined as being between 14 and <18 years, is not generally considered capable of making medical decisions on his/her own (Hartman, 2002). However, once an adolescent is an adult, they are responsible for making medical decisions. Despite recurrent VOEs in the adolescent and young adult (AYA)

population, it is unknown at what level of involvement AYAs are included in the healthcare decision making (HCDM) process surrounding treatment of VOE. Autonomous HCDM should evolve to become an independent activity as the AYA transitions to adult health care between the ages of 18 and 21 years, dependent on clinic policy. This is especially important with chronic diseases, as with SCD, which may include frequent occurrences of complications and the need to understand the impact on the body and treatment needs. While it is not known how or whether an AYA may be involved in HCDM, it is known that not all parents allow the adolescent to participate in the HCDM process, despite the adolescent being developmentally ready to participate (Miller & Drotar, 2007).

Decision-making capacity is defined by the processing of information and comprehension by the AYA with an understanding of the disease and proposed treatment information as postulated by the healthcare provider (Ruhe et al., 2015). Decisions take into account available options resulting in selection of an option believed to provide the desired outcome (Byrnes, 2002). Collaborative HCDM involves the parent and AYA taking into account activities such as AYA advice-seeking and parent-adolescent negotiation (Miller, 2009). It is unknown how or if parents teach AYAs with SCD to make decisions regarding health care for VOE. Adolescents and young adults tend to seek adult input less often as they age, relying on past experience to assist in HCDM (Byrnes, 2002). It is further noted that one must be able to constructively weigh options such as comparison of treatments and understanding of risks, benefits, and potential outcomes. The purpose of this study was to explore the thought process of adolescents and young adults with sickle cell disease, as well as their parents/legal guardians in healthcare decision making when considering emergency department treatment for acute vaso-occlusive pain episodes. The

aim of this study was to identify and compare factors that influence the parent/legal guardian's and AYA's healthcare decision-making when deciding to seek care from the emergency department for VOE in SCD. A secondary aim was to compare and contrast parent or legal guardian and AYA perceptions about the AYA's ability to seek emergency department care for VOE in SCD.

### **Methods/Methodology**

#### **Design**

This study used a qualitative descriptive design, with the Dialogical Self Theory to provide a lens through which to understand and explain the HCDM phenomenon in AYAs with SCD. (Colorafi & Evans, 2016). Individual perspectives of two individuals of the same event provide two narratives. Thematic analysis methods were used to analyze semi-structured interview data of parents/legal guardians and their AYA with SCD. Qualitative methods were chosen to help elucidate the unknown factors that influence the collaborative decision-making process between parents /legal guardians and AYAs when considering emergency department care for acute VOE.

#### **Theoretical Framework**

Dialogical Self Theory explains the self in relation to the thought process with HCDM. The self is composed of two parts: one part of the self is located within the mind and the other part extends to the environment (Hermans, 2012). Dialogical Self Theory (Figure 4) was used in this study to examine internal conversations related to the HCDM in AYAs with SCD.

Mikhail Bakhtin added the concept of the involvement of multiple voices to inner dialogue as cited in Hermans (2001). These multiple voices are portrayed as thoughts

within the mind of the person making the decision and may potentially include parents/legal guardians, physicians, and community. Collective voices or thoughts are influenced by health care providers, jargon, culture, social circles, teachings, and other such entities. As a result of this community of voices, the decision makers may have conflicting thoughts about their decision. The culture of medicine and the voice of the healthcare provider may influence the patient-self to seek care. Thoughts about what healthcare providers taught and how the AYA was treated in the hospital may work to dominate the discussion with the parent/legal guardian. These voices, both as part of the self and as belonging to me, become a story, agreeing and disagreeing, that results in the development of a solution to the discussion at hand based on possessed knowledge and experience. Exploration of this process related to visiting the emergency department for a VOE due to SCD aided in understanding how AYAs process thoughts related to healthcare decision-making and indicates needed teaching prior to transition to adult care.

### **Study Setting and Recruitment**

A convenience sample of parent/legal guardian and AYA cohorts was drawn from AYAs seeking care for SCD from a city and metropolitan area children's hospital in Georgia between June 2023 and December 2023. The hospital has an SCD specialty clinic and provides SCD care in the emergency department for VOE in children and AYAs to age 21 years.

The AYA and parent or legal guardian were approached by a member of the emergency department research team and provided with a study flyer containing a brief description of the study and contact information for the first author. Permission to be contacted via phone was obtained and contact information was shared with the first author

using a HIPAA protected SharePoint folder. Thirty-seven AYAs and their parent/legal guardians were referred. Parents/legal guardians were contacted by the first author, and the study was explained to them in detail (Appendix C). As a foster parent without the ability to consent for research, one was ineligible, and twenty-seven denied an interest in this study. Nine parents/legal guardians and seven AYAs were consented or assented. The interview with each participant was then scheduled. No payment or compensation was offered for participation in this study.

### **Inclusion and/or Exclusion Criteria**

Adolescents and young adults approached in the emergency department were being treated for acute VOE and ranged in age from 14 to less than 22 years of age. They had a diagnosis of SCD, no genotypes were excluded. The parent/legal guardian of the AYA present in the emergency department approached was 18 years of age or older. All participants were fluent in English. Participants were limited to those self-identified as African American. No exclusion was based on sex.

### **Data collection & Ethical considerations**

Children's Healthcare of Atlanta Institutional Review Board and the University of Missouri Institutional Review Board approved this qualitative, descriptive study to obtain data from parents/legal guardians and AYAs regarding HCDM.

The first author used a semi-structured interview guide to conduct the (appendixes D and E). The interview guide was developed using sickle cell literature related to parent and AYA HCDM related to seeking care for complications of chronic illness, including SCD and diabetes (like SCD diabetes requires daily HCDM). Dialogical Self Theory and the investigator's experience working with the sickle cell patient population over the past

14 years influenced interview guide development. Based on this information there were four domains of questions developed. Participants' responses to initial questions prompted additional questions and probes were developed to solicit additional details. Interviews lasted 15 to 25 minutes. The first author wrote field notes during the interviews and interviews were recorded using a digital recorder.

### **Data Analysis**

Reflexive thematic analysis (Rolfe, Ramsden, Banner, & Graham, 2018) was used to analyze data using MAXQDA 2022 for data management. Reflexive thematic analysis allowed pattern identification and allowed for both inductive analysis which helped identify themes that were derived directly from the data itself without relying on the Dialogical Self Theory framework, and deductive analysis which analyzed and confirmed the theoretical constructs consistent with the guiding theory. Use of this method allowed for examination of participant experiences while deeply exploring their meanings (Braun & Clarke, 2006; Braun, Clarke, Hayfield, Davey, & Jenkinson, 2023; Byrne & quantity, 2022). Inductive coding collected phrases, segments, and words from the participant perspective. Deductive codes were developed a priori based on Dialogical Self Theory with attention to the primary research questions. Inductive coding was then incorporated using the raw data to add understanding of what participants thought regarding HCDM for emergency room treatment of their VOE (Azungah, 2018). Coding was organized to develop themes (Figure 5). Analysis proceeded using the six steps in thematic data analysis which included 1) familiarization, 2) coding, 3) generating themes, 4) reviewing themes, 5) defining and naming themes, 6) writing the report that captured the essence of the analysis (Braun & Clarke, 2006).

## **Findings**

Nine parents/legal guardians agreed to participate in the study (Table 3); eight had AYAs under the age of 18 years (Table 4) and all agreed to allow their AYA to participate. Two parents/legal guardians did not answer the phone despite five attempts and two texts. This resulted in seven AYAs participating in the study, including one over the age of 18 years. All but the adult AYA were still in school and four AYAs were male. There were eight female parents/legal guardians. Demographic data are presented Tables 3 (AYA) and 4 (parent/legal guardian).

Four themes emerged to elucidate the aims of the study. The experiences of the AYA as described by AYAs and their parents/legal guardians in HCDM regarding emergency department care and the manner in which they reached that decision were as follows: (1) Knowledge acquisition through learning activities or identification of information or evidence required to care for self and make medical decision; (2) Thought process in HCDM through consideration of reasons for emergency department care; (3) Discussion to seek VOE care in the emergency room; and (4) Reasons it is the primary responsibility of the AYA to make the decision to seek emergency department care (Table 5 and Table 6).

### **1) Knowledge acquisition through learning activities or identification of information or evidence required to care for self and make medical decisions**

Six AYAs described learning activities as discussions with mom/parents about SCD and VOE care. They described the development of an understanding of how to treat a VOE through repeated demonstration by the parent/legal guardian throughout the years

as the AYAs experienced pain. They stated that as they had repeated VOEs and as they got older, they began to discuss the home treatment plan and initiate this plan themselves. Despite learning what to do, five AYAs asked their parent/legal guardian at some point if there was anything else they should be doing.

*“I learned from my parents. They just told me about everything, What it is, how to treat my pain and that I have to take medicine every day.” (ID#101)*

Adolescents/young adults described parents/legal guardians as being involved in their HCDM learning. This was described as learning through example and replicating parent treatment decisions. One participant stated he learned from caring for himself, trying things he learned from his parents, noting whether they worked. Four stated they learned about SCD from healthcare providers when they were younger or during discussions at clinic visits while three stated they did not learn about HCDM from healthcare providers but rather, from their parents/legal guardians.

*“ Like when I had pain they always made me drink a lot of water while I was sick and Waking me up to take my medicine around the clock. So as I've gotten older I just did it by example”. (ID#110)*

Six parents/legal guardians described themselves as monitors for pain management at home. They stated that they would review and sometimes become involved with what the AYA has done to alleviate pain at various intervals throughout the pain episode and discuss what the AYA might try next. They also said that the AYA usually starts treating the pain themselves, often before the parent/legal guardian is aware they are in pain. Two parents/legal guardians stated they monitored/administered the opioids to the AYA, but the others stated the AYA can take it on their own.

Except for one, parents/legal guardians described healthcare providers involvement in educating AYAs regarding HCDM for VOE as discussions during clinic visits, noting that it is important that everyone is involved in teaching the AYA. One parent also described classes that are offered to AYAs during transition as being helpful in AYA understanding of VOE care and medication management. One parent/legal guardian took the AYA to conferences and an SCD support group to assist in their learning regarding SCD and its management.

**2) Thought process in decision making includes reasoning for emergency department care**

The degree of pain did not play an important role in the decision to go to the emergency room as the pain was often severe at home. Rather, inadequate response to symptom management was a determining factor. The thought process described by AYAs and parents/legal guardians when considering emergency department care was focused on the treatment provided for VOE and the possibility of the AYA obtaining pain relief. Upon arrival at the emergency department AYAs were quickly seen, given intravenous fluids and either nasal spray or intravenous pain medication. Relief was not always immediate, and staff were attentive to the need for multiple doses of medication and possible need for admission to inpatient care.

*“I was just like I need to go to hospital because like the medicine is not working and I can't hold no food down or no drink or water.” (ID# 102)*

Neither the AYAs nor the parents/legal guardians considered other factors such as transportation in the decision-making process. As assessment for stigma, all participants in each cohort were asked how the AYA was treated by staff and if there was any indication

it was believed the AYA was drug seeking. All stated they were treated well. One parent (ID #211) said she heard that adults were treated like drug seekers in the adult setting, but their AYA was never treated that way in the pediatric setting. Despite admission to inpatient care on an average of 9.86 (SD 4.38) occurrences during the year prior to the interview, only one cited potential hospitalization as a factor in decision making. One AYA stated she was stubborn and knew she would be admitted so she delayed going to the hospital. She also stated she was a 'hard stick' and so delayed seeking emergency department care. Three other AYAs or their parents/legal guardians stated that it was difficult to access a vein and so the AYA would be given nasal opioids initially and then intravenous access would be obtained. Two parents/legal guardians did state they called the SCD clinic or on-call healthcare provider to let them know what was going on and affirm the need to go to the emergency department. One parent/legal guardian called the emergency department to prepare them for the AYA presentation for a VOE.

### **3) Discussion to seek VOE care in the emergency room between the AYA and parent/legal guardian**

Discussion was most often initiated by the AYA due to an ineffective response to home care but in two cases the parent/legal guardian intervened first, stating that the AYA needed to go to the emergency department. They both stated that they know their AYA well enough to be able to look at the AYA and recognize that home care is not working. This was also described by two AYAs as the initiation of discussion regarding emergency department care.

Discussion regarding visiting the emergency room was described by parents/legal guardians as confirmation that the AYA had appropriately managed the VOE at home

without relief, as described by five parents. While this discussion occurred, the parents/legal guardians felt the ultimate decision was AYA's right because they knew the severity of the pain and what they could tolerate. It was also noted that the AYA had dealt with enough pain over the years such that they could make the decision.

*"We always leave it to her because usually when we ask her she doesn't want to go. So we don't, we don't usually initiate, it's we let her let us know. She'll just tell, she'll just let us know like oh I can't deal with the pain anymore. It's too bad. So I need to go in."* (ID# 210).

**4) AYAs are the ones experiencing the pain and are old enough to understand the implications of emergency department care**

All AYAs expressed the belief that it is their body and thus they should make decisions about treatment. They explained that they have been experiencing VOEs for most of their life and are aware of the severity of pain they are having when considering emergency department care. Five described that they knew they could no longer tolerate the pain and felt the need to go to the emergency department. They all described VOE care initiated by themselves and discussed it with their parent/legal guardian at home. They noted that home care was not lessening pain intensity or length of VOE, and that pain was often escalating.

Parents/legal guardians agreed with the AYAs, that it was the AYA's body, and thus they should be the person to decide when they can no longer tolerate the pain and require emergency department care. Parents/legal guardians described the home care initiated and stated that it was not adequately alleviating the pain, expressing agreement with the AYA that emergency department care was required.

AYAs also described their level of maturity as a factor in HCDM. They stated they were in their teens with three stating they were almost adults. Four described an understanding between the ages of ten and thirteen of their ability to connect the type of pain they had and their ability to care for it at home versus emergency department care. Adolescents and young adults also described themselves as being old enough to understand their pain and how to treat it and how it responded to treatment.

Five parents also agreed that children between the ages of ten and eleven should begin to participate in HCDM because they can begin to understand pain. Seven parents further stated that AYAs are old enough to make decisions alone or in unison with their parent/legal guardian regarding their pain.

*“If they are old enough and mature enough to understand. Most adolescents I know, being in different groups and chatting. When they have a chronic disease, they are a lot more mature and can understand and make those decisions. (ID# 201)*

## **Discussion**

The results of this study capture the individual perspective regarding HCDM of AYAs during a VOE and stresses the importance of early education to assist in the management their chronic disease. These findings have the potential to enhance care of AYAs and their transition to adult healthcare management of their chronic disease.

The literature reports that AYAs under the age of 18 are unable to legally make independent medical decisions and those over 18 may include their parent/legal guardian in the process (Hartman, 2002). The AYAs in this study, despite six of them being under the age of 18 years, expressed a desire to make the decision regarding emergency department care for VOE. They cited an understanding of their body and their ability to

tolerate pain while being at home versus the need to seek care. Parents/legal guardians noted that AYAs were able to understand their pain tolerance and should be involved in the decision to seek emergency department care. Despite their age, all AYAs in this study expressed a desire to be the primary decision-maker and provided plausible reasons for such beliefs.

### **Knowledge acquisition through learning activities or identification of information required to make medical decisions**

Manwani et al. (2022) reported interventions for transition to adult care in SCD designed to assess readiness for transition and goals of AYAs while working to remove barriers and provide important knowledge about the disease and symptom management improved transition readiness. This is essential to the continued care of AYAs in the adult setting as they become responsible for self-care and HCDM.

Adolescents and young adults in the current study primarily considered the relief from VOE pain they would receive when seeking emergency department care as the motivator in their decision. Other factors that may impact the acceptance of emergency department care in young adults were not present in AYAs consideration for emergency department care in the current study but had been noted in a past study by Bulgin et al. (2018) which explored reasons young adults avoided seeking emergency department care for SCD VOEs. Bulgin et al. (2018) noted AYAs often stated they either learned to do so through experience or were told by healthcare providers or caregivers to avoid the emergency department. Emergency department treatment in a pediatric setting was reported by AYAs in Bulgin (2018) study when asked as excellent. Conversely this was not true for young adults in an adult care setting, as they felt they were often perceived as drug-

seekers (Bulgin et al., 2018). They also noted other reasons for young adult avoidance of emergency department care in AYA decision making. These included transportation, intravenous access issues, lack of insurance, and concern for possible in-patient admission. Parents/legal guardians, as with AYAs in this study, expressed a desire to seek pain relief for the AYA, stating that the AYA was well treated, and their pain was adequately addressed.

Dialogical self-theory (Wray-Lake et al., 2010) was used to examine internal conversations related to the decision-making process related to decisions made during the past year using an adolescent decision-making questionnaire. Exploration of this decision-making process aided in understanding how AYA process thoughts related to HCDM and indicate needed teaching prior to transition to adult care. A decision takes into account available treatment options and behaviors related to selection of the option believed to provide the desired outcome (Byrnes, 2002). Decisions regarding home care included both non-pharmacological and opioid treatment. AYAs reported self-care they thought would help based on prior care provided by parents/legal guardians for past VOEs. When this care did not work, they considered emergency department care.

Parents/legal guardians and AYAs expressed the belief that AYAs were primarily responsible for HCDM when considering medical VOE care. While chronological age is a consideration in transition to adult care, research indicates demonstration of self-care is more important in AYA readiness to assume self-care responsibilities (Reed-Knight, Blount, & Gilleland, 2014). Autonomy also factors into the ability of the AYA to take responsibility for their health as they master tasks important to their daily life (Reed-Knight et al., 2014).

Our study found that AYAs felt they were able to make decisions because they understood their own body and how they responded to healthcare management. In a study exploring AYA involvement in cancer treatment decision making it was found that they felt they had enough experience with the disease to be able to understand their healthcare needs (Pyke-Grimm, Franck, Halpern-Felsher, Goldsby, & Rehm, 2020). Further, AYAs in this study learned about their care through observation of healthcare providers and other patients. Knowledge acquisition allowed them to determine healthcare preferences and respond to their own needs.

Parents/legal guardians in this study were primarily approached by the AYA to initiate discussion of the need to visit the emergency department to manage their pain. Adolescents and young adults noted that discussion and demonstration of VOE management by parents/legal guardians over the years led to an understanding by the AYAs of the best way to treat their VOE at home and when to consider emergency department care. These findings indicate that more research is needed related to parent-adolescent communication regarding VOE to ascertain the best way for parents and AYAs to interact with a precursor to the development of intervention for younger children and parents regarding home and medical care. It also indicates that parents may need to start conversations about self-care prior to adolescence to prevent some negative care discussions and increase adolescent autonomy.

### **Limitations**

This study had several limitations. This study was limited by the ability to recruit at only one hospital emergency department given that the other two hospitals in the system did not have a research team capable of approaching the patient and family during an

emergency department visit. Second, some parents/legal guardians and AYAs stated they were not comfortable interviewing over the telephone. Fourth, it is unclear how well children/adolescents of various ages understand their disease and the level of their self-care knowledge and their level of decision-making skills. Finally, due to telephone interview scheduled at a time different than consenting (only one parent was available to interview at time of consenting), some potential participants did not answer the telephone at the scheduled time or respond to voicemails or text messages and were thus lost to follow-up. Finally, this study explored a small cohort of African American AYAs and their parents/legal guardians with heterogeneity in their diagnosis, history of chronic VOE complications, and prior emergency department experiences in a controlled setting, all factors which may have influenced the perspectives of their HCDM process.

### **Recommendations for Further Research**

Future research in sickle cell HCDM could focus on development of patient-centered decision aids to help individuals, and their caregivers weigh the risks and benefits of different treatment options and when to elevate healthcare seeking to hospital level care in a culturally sensitive framework.

Another area for future research consideration would be to explore health literacy and its influence on outcomes and shared decision making. Discussions between parents/legal guardians and AYAs have been explored to a limited degree in some diseases, most notably diabetes. While discussions regarding healthcare within these two groups are important, HCDM by younger children with SCD and other chronic diseases should include their understanding of disease and self-care knowledge should be explored. Mixed-method research should include health literacy and self-efficacy regarding SCD while

exploring the HCDM process for various treatments to prevent and treat complications and new treatment options for SCD. An exploration of self-care and HCDM to understand the thought process and factors considered in decision making while exploring the individual's understanding of the disease and treatment options may lead to better development of teaching plans for patients and caregivers.

### **Conclusions**

AYAs with SCD describe HCDM regarding treatment of VOE when seeking emergency department care as primarily their responsibility. While they discuss it with their parents, they feel they know their body best and are old enough to make such decisions. Despite reviewing a plethora of articles discussing transition program planning and activities in a variety of chronic diseases, the first author (DR) was unable to find any transition programs that specifically addressed HCDM. Decision-making is often a shared activity between the healthcare provider and the patient for initiation of treatment or intervention. However, it can be an independent activity when addressing daily healthcare activities for chronic disease, such as diet and exercise in diabetes and pain management in SCD. It is imperative that children and AYAs receive appropriate education to make decisions and are actively involved in the process.

Figure 4 Dialogical Self Theory

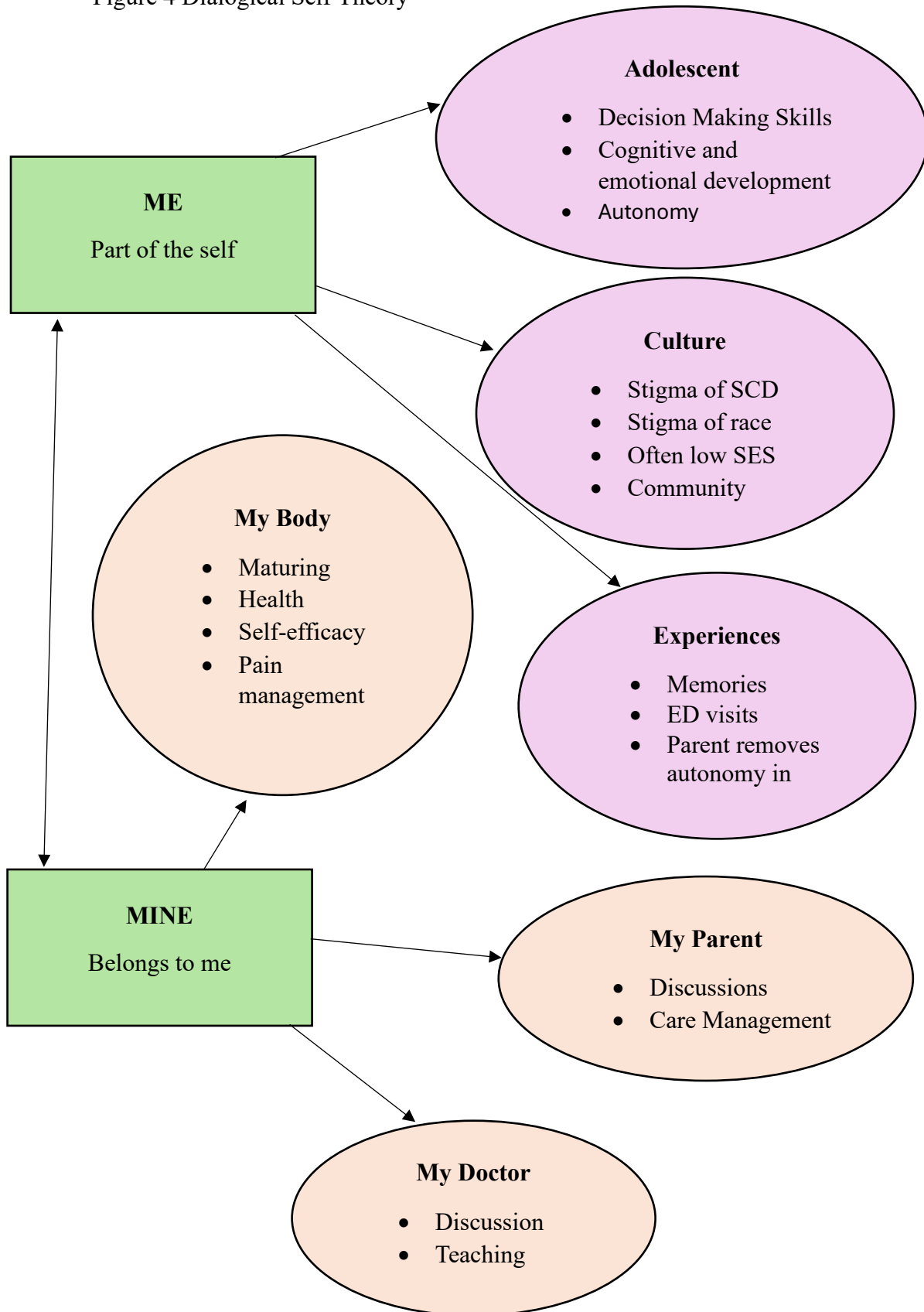
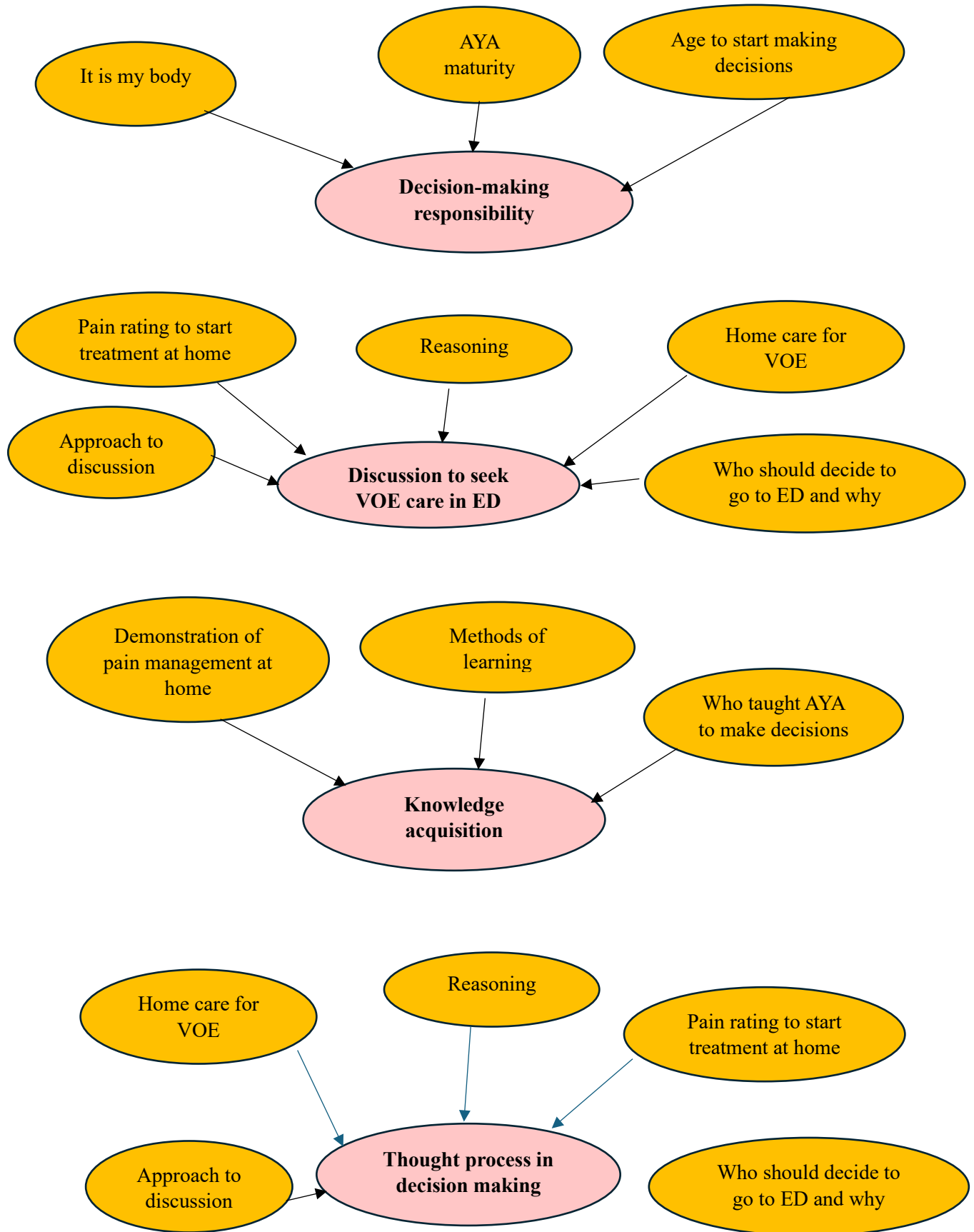


Figure 5. Theme Development



**Appendix C** 

## Phone script

My name is Diana Ross, I am a PhD nursing student working on my dissertation. This study is intended to explore your experience planning to go to the emergency department to seek care for your (your adolescent's) pain episode. It involves an interview with you and your parent/adolescent. We will do this interview over the phone and I will record it. The interview will take 30 to 45 minutes. I ask that you are in a room where no one else can hear your conversation so that the information remains confidential. I will also collect demographic information, including age, gender, year in school/employment status, race and ethnicity. There is a possibility that others may see your interview transcription. The interview and demographic data will be de-identified, meaning it will only have an ID number on it, not your name and will be stored on a password protected share drive at Emory University. If you agree to participate, we will set up a day and time to do the interview that works best for your schedule.

**Appendix D. Semi-structured Interview Guide Adolescents**

So first, I just want to get to know a little about you.

16. What grade are you in this year at school or have you graduated?
  - a. (if graduated) are you in trade school or college or do you work?
    - i. (if so in school) What are you studying?
    - ii. How do you like it?
    - iii. What was your favorite subject in high school?
  - b. (if graduated and work) What do you do for your job?
    - i. What is your favorite part of your job?
    - ii. Tell me about your typical workday.
    - iii. What kinds of things do you do when you are not at work?

Thank you. I want you to think about your most recent visit to the emergency room for a pain episode and treatment. I am going to ask you some questions about how you and your parent (s) decided to go to the emergency room.

17. How do you learn about sickle cell disease?
  - a. Parent teaches
  - b. Nurses, doctors
  - c. Reading
  - d. Internet
  - e. Other
18. Tell me about your last pain episode.
  - a. Triggers
  - b. Describe the pain
  - c. How would you rate the pain on a scale of zero to ten with zero being no pain and 10 being the worse pain you can imagine?
  - d. Did you talk to your parent about how to treat your pain at home?
  - e. Tell me about what you both said
    - i. What were you thinking about when you were talking about treating your pain?
    - ii. Other pain episodes
    - iii. Ways to treat you pain
    - iv. Ways you treated your pain in the past
    - v. Other
    - vi. What did you decide to do about your pain?
      1. Home pain plan
      2. Meds
      3. Water
      4. Heat
      5. Rest

## 6. Other

- f. Who made the decisions about how to treat your pain at home?
19. When that treatment did not work, what did you and your parent talk about?
  - a. Tell me about your pain at that time.
20. What were your thoughts when you were talking with your parent about your pain?
21. When did you decide to go to the emergency room?
22. Tell me about your pain at that time.
  - a. Describe
  - b. Rate
23. What were you thinking about when you were deciding to go to the emergency room?
  - a. Tell me what you and your parent talked about when you were making this decision.
  - b. Who made the decision to go to the hospital?
    - i. Tell me what you and your parent talked about when you were deciding
    - ii. What were you thinking about?
      1. Emergency room pain plan
      2. Past emergency room visits
      3. Doctors' advice
      4. What would happen when you went to the emergency room
24. Tell me about your visit to the emergency room
  - a. How long did you have to wait to be seen
  - b. Who described your pain and treatment at home to the medical staff
  - c. What type of treatment did you receive
  - d. How did the staff treat you
25. Tell me about some of the health decisions you make regarding your sickle cell disease?
  - a. Tell me about what you and your parent talked about when you were deciding how to make this decision
26. Tell me who you think should decide how you treat your pain at home
27. Who should decide when you should go to the hospital for your care?
  - a. How old should a child or adolescent be when starting to make decisions to the ED for VOC
28. How do you want to learn to make decisions about your care?
  - a. Parent teaches
  - b. Doctors and nurses teach
  - c. Reading
  - d. Other
29. Why do you think you have enough information to make decisions about your sickle cell disease?

30. Do you want to make decisions about your sickle cell disease and why or why not

### **Appendix E. Semi-structured Interview Guide Parents**

So first, I just want to get to know a little about you and your adolescent.

17. Tell me a little about your adolescent or young adult child.
  - a. If in school, what grade are they in?
18. Do you work outside the home?
  - a. What type of work do you do?
19. Tell me a little about a typical day for you.
20. Is your adolescent or young adult still in school?
  - a. Do they work?
  - b. What is a typical day for them?
  - c. Tell me about what happens when your adolescent or young adult has a pain episode.

Thank you. I want you to think about your adolescent's most recent visit to the emergency room for a pain episode and treatment. I am going to ask you some questions about how you and your adolescent decided to go to the emergency room.

21. Tell me what that pain episode was like for your adolescent.
  - a. Probes
    - i. Triggers
    - ii. How does he/she describe the pain?
22. Tell me about their last pain episode.
  - a. How did they describe it?
  - b. Do you know what triggered it?
  - c. What did you do at home for the pain?
    - i. Who decided how to treat the pain?
    - ii. What were you thinking about when you were making this decision?
      1. Ways to treat the pain.
      2. Home pain plan
      3. What your adolescent wanted to do to treat the pain
      4. What worked in the past to treat the pain?
23. Tell me about the things you were thinking of during this conversation to treat the pain.
  - a. How did you go about making that decision?
  - b. Who was involved in making the decision?
    - i. Tell me about the conversation you had with them to make this decision.
24. When the initial treatment did not work, what was the discussion like between you and your adolescent about further treating the pain?
  - a. Opioids, other treatments.

- i. Tell me about your understanding of the pain at that time.
- 25. What were your thoughts when you were having this discussion with your adolescent?
- 26. Tell me about your visit to the emergency room.
  - a. How long did you have to wait to be seen?
  - b. What type of treatment did you receive?
  - c. How did the staff treat you?

Now I want to talk about when your adolescent's pain episode got to the point that you and your adolescent decided that at home treatment was not working.

- 27. Tell me your understanding of your adolescent's pain at that time.
- 28. What were our thoughts when you were considering taking your adolescent to the emergency department?
  - a. Tell me about your discussion with your adolescent when considering going to the emergency room.
  - b. Tell me how the final decision to go to the hospital was made.
    - i. Who made the decision and why?
  - c. What were your thoughts when making this decision?
    - i. Emergency room pain plan
    - ii. Past emergency room visits
    - iii. Doctors' advice
    - iv. What would happen when you went to the emergency room
    - v. Transportation
    - vi. Childcare
- 29. How has and is your adolescent learning about sickle cell disease?
  - a. Parent teaches.
  - b. Doctors and nurses teach.
  - c. Reading
  - d. Other
- 30. Tell me about some of the health decisions your adolescent makes regarding his/her sickle cell disease.
  - a. Who should teach them to make decisions about sickle cell disease and healthcare?
  - b. How should they be taught to make decisions?
- 31. Tell me what you think of his/her decision-making abilities.
  - a. What do you think about how mature they are?
  - b. What do you think is their ability to make decisions about their healthcare?
- 32. How do you believe your adolescent wants to participate in making decisions about his/her sickle cell disease?

**Table 3**  
Parent/legal guardian demographics (N=9)

		Number (percentage)
Age	33-56 years	42.56
Gender	Female	8 (88)
Race	Black/African American	8 (88)
Ethnicity	Non-Hispanic	8 (88)
	Mixed race	1 (11)
Education	Graduated high school	4 (33)
	Some college	1 (11)
	Associate degree	1 (11)
	Bachelor's degree	4 (33)
	PhD	1 (11)
Employment	Full-time	6 (66)
	Unemployed	3 (33)
Marital Status	Single	4 (44)
	Divorced	3 (33)
	Married	2 (22)

**Table 4**  
Adolescent and young adult demographics (N=7)

		Number (percentage)
Age	16-19 years	16 (1)
Gender	Male	4 (57)
Race	Black/African American	6 (85)
	Mixed Race	1 (14)
Ethnicity	Non-Hispanic	6 (85)
Education	Grade high school	10 (1)
	Graduated high school	1 (14)
Employment	Part-time	1 (14)
Sickle cell phenotype	Hemoglobin SS	5 (71)
	Hemoglobin SC	2 (28)
Disease modifying therapy	Hydroxyurea	3 (42)
	Chronic blood transfusion	1 (14)
Medical care during year prior to interview	Emergency department visits	9.86 (4)
	Hospital admissions	6.86 (3)

**Table 5. Quotes**

Adolescents and young adults

Subject ID	Theme	Quote
101	Knowledge acquisition through learning activities or identification of information or evidence required to care for self and make healthcare decisions	The doctors just taught me how to like answer my pain. Like, um, the cold and hydrate.
102		I learned from my parents. They just told me about everything, What it is, how to treat my pain and that I have to take medicine every day.
105		Well, it was mostly like, I would have like a pain crisis, like at an early age, like high school and so my parents teach me like during when I had pain at home and whenever I'm like in a hospital or like the nurses would.
107		I think my mother had to explain to me why I was taking the pills and what they did for me. So it was my mom who taught be about sickle cell and then sometimes the doctors would offer like my pamphlets or little notebooks, kinda like coloring aspects of those little sketch books, you know for children, but a better idea of what your condition is while keeping it like entertaining for the children.
110		my parent, something like in my childhood my parents always did taught me. They would break down the science of sickle cell. They would also be like telling me how to deal with it in my body per se, like what triggers me, what could be a trigger, things to stay away from, Like when I had pain they always made me drink a lot of water while I was sick and Waking me up to take my medicine around the clock. So as I've gotten older I just did it by example.
	Thought process in decision making includes reasoning for emergency department care	

- 102 I was just like I need to go to hospital because  
like the medicine is not working and I can't hold  
no food down or no drink or water.
- 103 I was thinking it'll make me better. That's all  
105 Because I think about like when I am on the  
bridge up heading to the hospital, I think about  
what they can offer me that I haven't already  
done. That, anything they can offer me that I  
can't you know, get my hands on myself. Like  
any opioids narcotics. I know I have this at  
home. So I don't need to go to the hospital for  
this if I take this at home and it's still not  
working then I have to go. Um. you know,  
things like that.
- 110 At that point I was having pain for quite a  
while. So I was trying not to go to the  
emergency room and just treat it at home. But  
the pain wasn't letting up, it wasn't really going  
to go away and it was getting worse. So over the  
long, it was very close to me taking my meds  
again, thinking about that. well, I was thinking  
about like not wanting to go but like I have to  
go cause you know, all the needles and drawing  
blood, being on heavy medicine and all those  
things. It's a little nerve-racking
- Discussion to seek VOE  
care in the emergency  
room between the AYA  
and parent/legal guardian
- 101 I was just like, mommy, I can't do it no more. I  
gotta go.
- 103 I just go in the room and I start wearing down at  
home, I just think somebody don't have to say  
nothing like, that's how bad it be. So she can  
see, I don't have to tell her I need to go.
- 105 I tell her that it hurts. Yes, it's pretty much that,  
it's just I hurt. It's like I tell my mom that it  
hurts. I tell her I want to go to the hospital.
- 107 Um, I talk to my mom, and she always asks me  
like oh, have you taken anything. How long has  
it been? You know, just making sure that I  
plugged in my process prior to deciding to just  
head to the hospital. This the hospital is my last,  
she always just make sure that I've done

- 109 everything that can before we head to the hospital.  
She knew like, when I've already taken like a lot of medicine I tell her like I don't think the medicine is doing anything and I think I need to go to the hospital or it's going to get worse.
- Reasons HCDM regarding emergency department care for VOE is the primary responsibility of the AYA
- 101 It's my responsibility so I could keep up with my health and not know that I need to do what I need to do when I'm hurting.
- 102 I just think I should decide, me, cause I'm the one with the pain. And I'm good doing it
- 103 I'm old enough to do it myself. It's my body. I guess I just know what's best for like me.
- 107 Because at the end of the day especially with everyone has a different pain tolerance, you can't really be sure how much pain someone else is in. So I feel like I should always be like the last say in what happens in terms of going to the hospital.
- 109 Because I know my body and I know my pain and I know what can make my pain worse enough to go to the hospital.
- 110 I think it should be me because I think I am, well, I'm well above the age but yeah, I'm at that age where like I know my limit. So yes, I know my limits, I can speak up for myself, all those things.
-

**Table 6. Quotes**

Parents and legal guardians

Subject ID	Theme	Quote
201	Knowledge acquisition through learning activities or identification of information or evidence required to care for self and make healthcare decisions	<p>She was diagnosed at Birth. And I told her throughout the years, along with the Children's Healthcare XX staff. About how to try to focus on something else, dressing in layers. Making sure that we know how Georgia weather is. So if it's cool in the morning, go ahead and wear something, you know, to keep you warm. And then once it gets hot, you can be able to take it off.</p> <p>I get her tried not to do over-exert herself.</p>
202		<p>I teach him and the clinic where he goes. They do. And which they told me that they're going to have to start sending them to a class to let him know how he have to take his medicine for himself and because he's getting of age that he should be able to take his meds on his own. So I told him, you know, I just, you know, I won't be able to go in the classroom, but he got there said that they're going up, start showing him and teach them to ask him. Do you know if they had before? Why did we take his meds and stuff of that sort.</p>
203		<p>I just explained how they explained it to me, for him he's sick, you know, he have a crisis and the body sickled. I tried to explain to him what's going on and why is it happening? If you don't take your medication like you're supposed to, you know, this is what's going to happen to you. And you know, I don't know if he understand what I'm saying or where I'm coming from because we was going to send XXX to a class. So that, cuz he's got, you know, he's getting older. So they would talk to him this, you know, even though I'm explaining it to him, but they</p>

- could, you know, talk to him a little better than I can.
- 203 Both, the doctor and me (should teach about decision making) because I can only do so much, say so much, so I'm not a doctor to explain to him why his medicine is necessary? Why he need his medicine. You know, and all that stuff, you know.
- 205 I explain to him what the doctors explained to me and, you know, I understand, you know, the signs beforehand. So you can go try to kick the pain out. The earlier we catch it, you know, the earlier we can try to kick it out of your system, I think you know because he does have that urinary problem, especially just laying in that wet space is another Factor on top of that. And so It's just explaining to him the situation, what he needs to do and how he needs to handle himself. And it's not a hard, you know, I got to think about the situation. He understand this is his body and this is how the things he needs to do to keep himself together.
- 208 I taught him (how to make decisions) on kinda figuring out his level of pain that he can take before he makes a decision to go to the hospital.
- 209 So we did a lot with our community support groups. And that's pretty much just talking to other patients that are you know older and I've already kind of went through what they were going through and talking to other parents and that's pretty much how I like the basis of it. Now, it's just kind of branched into doing your own research and reaching out to other people that have went through the same thing and just talking to doctors. Well I don't know, I guess you kind of learn the trends and their lab work and their triggers that kind of thing.
- Thought process in decision making includes reasoning for emergency department care
- 203 I give the medication like they asked me to; I give him the oxycodone to see will it work. And if it doesn't work, that's when I called the doctor and let them know what's going on with him

- and they asked me how long it been since I gave him the medication and how long he's been in pain and I explain it to them. And so they tell me to wait and see if the oxycodone work and it doesn't so they tell me that's when I have to bring them in. So they go from there.
- 205 it's his pain reaction and then the fact that he's had his medication and it's still, he's still in extreme pain. After I could say after an hour or two, that if he's still in extreme pain and he can't feel that medication working for that he's in a full-blown pain crisis.
- 207 That's not my decision. I mean you can almost, she complain and how much she can take when it gets to a level where it's too much for her. I can't make that decision [00:04:40] for her. I'm just, I'm here to help her get better. I think about her comfort for anything else is not is deepness to me. I just want her to be comfortable. That's my only concern.
- 209 I mean I say there are definitely times, there have been a couple times where you're kind of discouraged from going because you're like we just went and sent he sent you home from the ER and I know you're not better. So like there's some of those I still take her.
- 211 I just ask and I talk to him about his pain, what he's going through and just because if it's if it's bearable with the medicine that he has at home, if he's not running a fever or anything and we at first will try to get treat him at home. There's the pain doesn't seem to get any better getting on the meds it easier if you just go to the emergency room.
- Discussion to seek VOE care in the emergency room between the AYA and parent/legal guardian
- 201 I ask her where's it at. You know, what do you need me to do. She just, I know she's tired, she want to be able to do the normal things that a teenage girl can do, but she can't.
- 203 Well, he comes to me and he let me know, you know, if he's in pain and stuff like that and he like you lay up on the bed and he crying and moaning. I say are you okay. He said, no, but

- 205 mom I need to go to the hospital. And I said, well, let's take the medication first and see how it works. See will it help you, you know, and I wait because, you know, we had to take him to take the oxycodone every, I think it's every 4 to 6 hours. But he still hadn't gotten any better because they don't want me to continue to give it to him like that, but I I try to give him the medication to see, will it work or calm down, calm the pain down or whatever, ease the pain, and it doesn't work. So I have to take him in. After I give him his pain medication because he'll say, you know, my back is hurt or whatever is hurting on him and I said well you know, take your pain medication and so he takes it and generally I don't hear him for a while and I go and check on him and he's still hurting to the extreme. I said, you know, did it not work for you and he would say no or shake his head no. I say do you need to go to a doctor? He says, yes. We always leave it to her because usually when we ask her she doesn't want to go. So we don't, we don't usually initiate, it's we let her let us know. She'll just tell, she'll just let us know like oh I can't deal with the pain anymore. It's too bad. So I need to go in.
- 210
- 211 I just ask and I talk to him about his pain, what he's going through and just because if it's if it's bearable with the medicine that he has at home, if he's not running a fever or anything and we at first will try to get treat him at home. There's the pain doesn't seem to get any better getting on the meds it easier if you just go to the emergency room.
- Reasons HCDM regarding emergency department care for VOE is the primary responsibility of the AYA
- 201 If they are old enough and mature enough to understand. Most adolescents I know, being in different groups and chatting. When they have a chronic disease they are a lot more mature and can understand and make those decisions.
- 202 Basically, he's old enough now to where he can let me know if he needs to go or not.

- 203 He needs help cause I have to, I tell him as you get older you're going to have to make decisions, make sure you take your medication without me, you will have to make sure you know, I said because he's 16 and he soon will be 18 and he will have to make these decisions on his own. He gotta understand why he take his medication and what it's for even though I tell him, but he got to make those decisions to make sure he takes his medication.
- 207 I think it depends on every child. I would say probably about 10.
- 209 Yeah, I mean they can help to decide at about 9 or of 10. I trust her judgment when it comes to deciding, she is 16 so she has been helping to decide for a couple of years.
- 211 Um, around, to me he's getting to the age now where he can make those, he's to the age where he knows how he feels so he's getting to the point where he can make those decisions when he needs to and doesn't need to.
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## Chapter V

The purpose of this qualitative descriptive research study was to explore the healthcare decision making (HCDM) process of adolescents and young adults (AYAs) when considering medical care for an acute vaso-occlusive episode (VOE) of pain in sickle cell disease (SCD). The results of this study identified four factors that influenced AYA's HCDM. These included 1) knowledge/understanding of how to treat their pain episode, 2) inadequate response to home pain management, 3) joint discussions with parent/legal guardian and 4) AYA's knowledge of what has helped in the past and how they feel when considering emergency department care. Additionally, both the parents/legal guardians and the AYAs expressed a belief that the AYA was capable of making the decision to seek emergency department care due to their age and an understanding of their body and its response to pain management attempts in the home setting. This study is the first to explore the decision-making process by AYAs with SCD and their parents/legal guardians when treating a VOE. This study took place at a hospital in a geographical area with a large population of patients with SCD and has electronic medical records containing a pain plan designed by a hematologist that is accessed and applied following presentation to the emergency department.

Knowledge acquisition is the first step in the HCDM process regarding self-care for VOE in the home setting and follow-up consideration of emergency department care. The AYAs in this study described minimal formalized education regarding SCD at an early age with an increase during preparation for transition though they denied they were taught HCDM when preparing for transition. They noted that most of their knowledge regarding home management of VOEs, SCD, and HCDM was obtained through observation and interaction with their parent/legal guardian during a VOE. There was significant lack of standardized methods of education after the provision of coloring books and handouts at approximately the age of five to seven years. Participants further

went on to identify that there were very non-descript conversations during outpatient visits with parents/legal guardians, children, and adolescents in relation to SCD and its management until the a formal program of transition began in adolescence.

Participants in this study indicated that they believe children should begin assisting in treatment decisions for VOE as soon as they understand how their pain responds to treatment in the home setting as well as in the emergency department. The age in which the participants, both AYAs and their parents/legal guardians, believed this understanding occurred was approximately 10 years of age. This indicates that a formal and continuing program of education for children from a young age regarding SCD, the treatment of VOEs in the home setting, and HCDM is required in the outpatient clinical setting. Education regarding child and AYA inclusion in healthcare management for SCD and HCDM regarding treatment options needs to also include education focused on the parents/legal guardians on techniques to include the child and AYA in this process.

Ideally, patient education should begin as soon as the child is able to comprehend that they have a chronic disease; education should include signs of well-managed health as well as indications of complications. It should culminate in a program of transition preparation beginning in adolescence and continuing until the AYA physically transitions to the adult care setting. There must be reinforcement of what was initially taught about SCD in early childhood as the child matures and can begin to understand the impact of what is being taught on self-care, medication use, and HCDM on their health status. and providing contact with local support groups. This teaching program should be standard policy and designed to be provided by primary healthcare providers and nurses during routine outpatient clinic visits.

An educational component of HCDM should be included in the clinical setting as part of the SCD management treatment plan. It should focus on the relationship of treatment to symptoms

of disease, timeliness of decisions, projected outcome(s) of treatment, and impact of personal values such as religion or fertility. This program should discuss acute care needs as well as daily and regimented management of disease, such as with medication or chronic blood infusion. Such a developmentally appropriate program of education should start as soon as the child is able to understand the impact of treatment options on pain management in the home setting and in the emergency department. Parents/legal guardians need to be involved in such discussions with factors elicited from them that are important to their HCDM efforts.

Adolescents and young adults in this study expressed a desire to be the primary decision-maker as a part of the self that includes an understanding of their body and its response to pain management interventions. Parents/legal guardians agreed that AYAs were primarily responsible for HCDM when considering medical VOE care, citing that the AYAs were old enough to begin to understand how their body responds to pain interventions. Decision-making capacity is required to make appropriate healthcare decisions (Miller, Drotar, & Kodish, 2004), so capacity needs to be established for the child or adolescent to make a voluntary decision. Children and adolescents involved in HCDM require the ability to understand their health and their healthcare needs and to communicate their treatment choices (Grootens-Wiegers, Hein, van den Broek, & de Vries, 2017). Children as young as 5 years of age understand language and may be able to verbally or nonverbally express their desires regarding healthcare. Cognitive reasoning related to healthcare needs, including risks and benefits of care may begin as early as 6 to 8 years of age but young children may not be able to fully apply reasoning to their decisions until the ages of 8 to 11 years. Children and adolescents should be included in conversations regarding healthcare and decision-making beginning at approximately 5 to 8 years of age with increased participation in

communication and HCDM as they demonstrate communication, understanding and reasoning related to their health, topics being discussed, and decisions to be made.

The thought process of AYAs in this study was focused on relief of pain. Past experiences related to VOE treatment at home and in the emergency department aided in the decision-making process as well as discussion with the parent/legal guardian. The parents/legal guardians expressed similar thoughts, resulting in agreement with the AYA regarding home care and the decision to seek care in the emergency department. Parents/legal guardians in this study were primarily approached by the AYA to initiate discussion of the need to visit the emergency department to manage their pain. Parents/legal guardians were receptive to the AYA bringing attention to their need to seek care in the emergency department. Discussion between the two entities included pain interventions applied in the home setting and the AYA's response to this treatment.

Bulgin et al. (2018) explored reasons young adults avoided seeking emergency department care for SCD VOEs, noting they often stated they either learned to do so through experience or were told by healthcare providers or caregivers to avoid the emergency department. While none of these factors were interwoven in the thought process of the AYAs or parents in this study, Bulgin et al. (2018) indicated they were important to young adults during HCDM to seek emergency department care. This implies that as the AYAs mature and become independent in self-care seeking, there are factors that should be considered and may not be pertinent as an adolescent under the care of their parent/legal guardian. Thus, factors such as transportation and insurance should be discussed with AYAs when providing education regarding emergency department care and HCDM.

These findings indicate that more research is needed related to parent-adolescent communication regarding chronic disease treatment to ascertain the best way for parents to interact

with their adolescents. It also indicates that parents should initiate conversations regarding self-care prior to adolescence to prevent some negative care discussions and increase adolescent autonomy. HCDM by younger children should include their understanding of disease and self-care knowledge should be explored with teaching provided by indicated level of maturity and understanding.

Most studies regarding HCDM in chronic disease, including SCD, focus on the parent/legal guardian decision-making. The need to study and intervene in the promotion of children and AYAs in HCDM is urgent as transition to adult care results in independent decision making by young adults. While the study of transition preparedness is increasingly being studied (Perry et al., 2017) there has not been any mention found by this author of HCDM preparation in transition programs.

Decision-making regarding medical treatment of AYAs may involve both the AYA and the parent/legal guardian. While adolescents under the age of 18 are unable to legally make independent medical decisions they need to be included in the process by both the parent/legal guardian and the healthcare provider. AYAs aged 18 years and older may include their parent/legal guardian in the process though this is not required (Hartman, 2002).

The National Heart, Lung, and Blood Institute (NHLBI) publishes guidelines for evidence-based management of SCD and recommends shared decision-making between patients and healthcare providers. Based on current research, these guidelines should include provision of methods of education regarding HCDM beginning as soon as the child can begin to understand the concept of medical care and its impact on SCD. A well-developed program of education to include continued and repeated presentation of age-appropriate materials for children through AYAs should be included. Guidelines for providers should provide methods of parent/legal guardian

inclusion in discussions during clinic visits as well as guidelines for child/adolescent education by parents in the home setting. As children develop their knowledge base regarding SCD they should be included in the shared decision-making process for healthcare management to encourage autonomy and ensure accurate knowledge is received.

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### Publications:

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2. Veludhandi, A., **Ross, D.**, Sinha, C., McCracken, C., Bakshi, N., Krishnamurti, L. *A decision support tool for allogeneic hematopoietic stem cell transplantation for children with sickle cell disease: Acceptability and usability study*. *JMIR Formative Research*, 2021. **5**(10): p. e30093.
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4. **Ross, D.**, Sinha, C., Bakshi, N., Krishnamurti, L. (2020) Educational needs of patients and caregivers living with sickle cell disease results in development of web-based patient decision aid. *Journal of Advanced Nursing Qualitative Research*, DOI: 10.1111/jan.14704
5. Sinha, C., Bakshi, N., **Ross, D.**, Loewenstein, G., Krishnamurti, L. (2020) Primary caregiver decision-making in hematopoietic cell transplantation and gene therapy for sickle cell disease. *Pediatric Blood & Cancer*. DOI: 10.1002/pbc.28749.

### Fields of Study:

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