

Health Care Transitions in Sickle Cell Disease

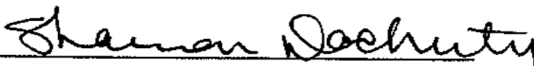
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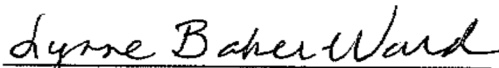
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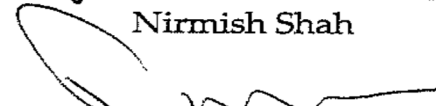

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Dissertation submitted in partial
fulfillment of the requirements for the degree
Doctor of Philosophy in Nursing
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ABSTRACT

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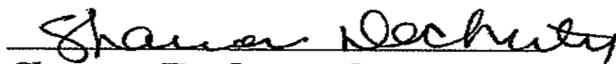
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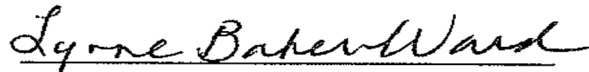
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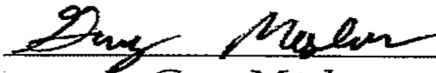
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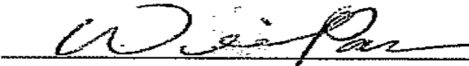
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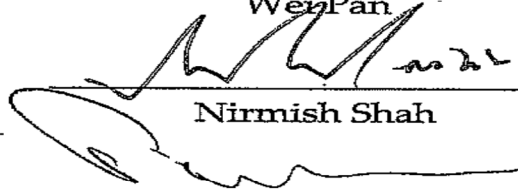
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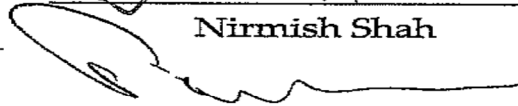
Gary Maslow



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Paula Tanabe

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Abstract

Over the past three decades, advances in the care for children with sickle cell disease (SCD) have increased their life expectancy and necessitated their transition to adult health care. However, there is a limited understating of health care transition in SCD and the impact it has on outcomes beyond perspective on and satisfaction with the process from the perspective of the adolescent and young adult (AYA) and their families. The purpose of this dissertation was to provide a better understanding of health care transitions in AYAs with SCD. This purpose was achieved through an examination of the state of the science on health care transition in AYAs with SCD, the challenges of shifting self-management from the parent to the adolescents with SCD prior to transition to adult care, and the patterns of health care utilization during transition and the factors associated with these patterns.

Findings from our integrative review indicated that few studies examined health care transition in AYAs with SCD. Most of the research on health care transition in AYAs with SCD focused on individual, family support, and the health care domain, yet results are inconclusive. For example, there is evidence that parents were heavily involved in the AYA's health care. Although this involvement was viewed as supportive by the AYA as well health care providers, it's not clear how parental involvement and the parent-AYA relationship can be utilized to facilitate health care transition for the AYAs with SCD. In addition, very few studies examined health care transition outcomes or examined how AYAs utilize the health care system to meet their health care needs during transition.

To further understand the parent-AYA relationship in managing SCD, we examined the shifting of management responsibility from the parent to the adolescent with SCD. Our findings indicated that shifting management responsibility is a critical and challenging process. The challenges that adolescents and parents face are adaptive-type challenges, rather than technical-type challenges. While technical-type challenges are easily fixed with technical work, more complex adaptive work is needed to address adaptive-type challenges, making the shifting of management responsibility more complex.

To understand health care transition in AYAs with SCD, we conducted a longitudinal examination of health care utilization for AYAs with SCD during transition to adult care and examined the factors associated with the different trajectory groups. Most AYAs in our sample had low clinic, hospital, and emergency department (ED) utilization trajectories. Few AYAs had high utilization in the clinic, hospital, and/ or ED. However, this group usually accounts for the highest costs of care. The sample mean emergency reliance scores were also below the cutoff point of 0.33 for high ED reliance. However, we did find evidence of increasing reliance on the ED compared to ambulatory care with increasing age indicating that a small group of AYAs with SCD might be at increased risk for high reliance on the ED. Individual, contextual, and transfer related factors were associated with the different health care utilization trajectory groups for AYAs with SCD. These factors varied between the different health care services indicating the complexity of health care utilization in AYAs with SCD and the factors influencing them. Receiving hydroxyurea was the only common predictor between higher clinic

and higher hospital utilization trajectory groups, while distance to the sickle cell center was the only common predictor between higher clinic and higher ED utilization trajectory groups. Depression, chronic pain, long-acting narcotics, and chronic transfusion at age 19 predicted higher hospital and higher ED trajectory utilization groups.

A major finding in this study was the association of several mental health conditions, including depression, with health care utilization group membership. This finding underscores the importance of routine screening and adequate management of mental health care conditions in AYAs with SCD, especially during transfer to adult care.

One goal of this dissertation was to describe successful transition. The majority of our sample has transferred to adult care. The mean age at transfer was 19 years. We also examined the continuity of care after transfer and the majority of participants who transferred to adult care had at least one additional encounter in the adult sickle cell clinic, and around 65% had more than ten encounters. Only a few (5.83%) had no adult encounters after transfer. As a result, we considered the vast majority of participants who had at least one encounter in the adult sickle cell clinic to have successfully transferred and integrated into adult care.

Dedication

To family, friends, mentors, and colleagues whose endless support made this work possible.

Contents

Abstract.....	7
List of Tables.....	15
List of Figures.....	16
Acknowledgements.....	17
1. Health Care Transition in Sickle Cell Disease: An Introduction.....	1
1.1 Overview of Sickle Cell Disease and Health Care Transition.....	1
1.2 Shifting Self-management in Preparation for Transition.....	4
1.3 Health Care Utilization.....	6
1.4 Theoretical Framework- Bronfenbrenner’s Bioecological Theory of Human Development.....	8
1.5 Purpose Statement and Research Aims.....	12
1.5.1 Aim one.....	13
1.5.2 Aim two.....	13
1.5.3 Aim three.....	13
1.5.4 Aim four.....	13
1.5.5 Aim five.....	14
2. Health Care Transition for Adolescents and Young Adults with Sickle Cell Disease: State of the Science Integrative Review.....	15
2.1 Theoretical Framework.....	16
2.2 Methods.....	17
2.3 Results.....	21
2.3.1 Description of studies.....	21

2.3.2 Health care transition outcomes.....	21
2.3.3 Individual domain.....	23
2.3.4 Family/Social support domain.....	29
2.3.5 Health care system domain.....	31
2.3.6 Additional findings.....	34
2.4 Discussion.....	36
2.5 Conclusions.....	46
3. Challenges in Shifting Management Responsibility from Parents to Adolescents with Sickle Cell Disease.....	47
3.1 Theoretical framework.....	51
3.2 Design and Methods.....	53
3.2.1 Study design.....	53
3.2.2 Participants.....	53
3.2.3 Data collection instruments and procedures.....	54
3.2.4 Data analysis.....	56
3.3 Results.....	57
3.3.1 Adolescent adaptive challenges.....	57
3.3.2 Adolescent adaptive work.....	60
3.3.3 Parent adaptive challenges.....	63
3.3.4 Parent adaptive work.....	69
3.4 Discussion.....	70
3.5 Study Limitations.....	76
3.6 Implications for Practice and Research.....	77

3.7 Conclusions	78
3.8 Acknowledgement	78
4. Health Care Utilization by Transitioning Adolescents and Young Adults with Sickle Cell Disease	79
4.1 Theoretical Framework.....	83
4.2 Methods	86
4.2.1 Databases and study population.....	86
4.2.2 Measures.....	87
4.2.3 Procedures.....	92
4.2.4 Data analyses.....	93
4.3 Findings	94
4.3.1 Sample characteristics.....	94
4.3.2 Health Care Utilization.....	102
4.3.2.1 Pediatric and Adult Clinic Utilization Trajectories.....	104
4.3.2.2 Hospital Utilization Trajectories.....	106
4.3.2.3 Emergency Department Utilization Trajectories.....	107
4.3.2.3 Coutilization Across Health Services.....	108
4.3.3.4 Emergency department reliance	109
4.3.4 Transfer to Adult Care and Continuity of Care	112
4.3.5 Factors Associated with Health Care Utilization Trajectory Groups.....	113
4.3.5.1 Factors associated with clinic trajectory groups.....	114
4.3.5.2 Factors associated with hospital trajectory groups	120
4.3.5.2 Factors associated with emergency department trajectory groups	125

4.4 Discussion.....	131
4.5 Limitations.....	137
4.6 Conclusion and Implications	138
4.7 Acknowledgement	139
5. Health Care Transition in Adolescents and Young Adults with Sickle Cell Disease: Future Directions	140
5.1 Summary of Findings.....	140
5.2 Limitations.....	146
5.3 Future direction for research and practice.....	148
References	151
Biography.....	169

List of Tables

Table 1: Database search terms and results.....	18
Table 2: Summary of articles and findings.....	37
Table 3: Study variables	87
Table 4: Sample characteristics.....	95
Table 5: Participants' disease complications, comorbidities, and outpatient medications.....	101
Table 6: Health care encounters by service type over the study period.....	103
Table 7: Fit indices for the regression models selected for the sickle cell clinic, hospital, and emergency department	114
Table 8: Parameter estimates and corresponding Wald test for clinic model	119
Table 9: Odds ratio estimates and corresponding 95% confidence interval.....	120
Table 10: Parameter estimates and their corresponding Wald test for hospital model.....	124
Table 11: Odds Ratio Estimates and corresponding 95% confidence interval	125
Table 12: Parameter estimates and their corresponding Wald test for hospital model.....	130
Table 13: Odds Ratio Estimates and corresponding 95% confidence interval	131

List of Figures

Figure 1: Prisma flow diagram.....	20
Figure 2: Mean number of vasoocclusive crises by age (ages 12-27) with 95% confidence interval	97
Figure 3: Mean number of acute chest syndrome episodes by age (ages 12-27) with 95% confidence interval.....	97
Figure 4: Mean modified sickle severity index at age 15 (left) & 19 (right).....	98
Figure 5: Mean number of blood transfusions by age (ages 12-27) with 95% confidence interval	100
Figure 6: Mean length of stay in days for all hospitalizations.....	103
Figure 7: Mean length of stay in hours for all emergency department encounters.....	104
Figure 8: Pediatric (left) and adult (right) sickle cell clinic utilization trajectories by participant age (ages 12-27).....	105
Figure 9: Hospital utilization trajectories by participant age (ages 12-27).....	106
Figure 10: Emergency department utilization trajectories by participant age (ages 12-27)	107
Figure 11: Mean emergency department reliance score by age (ages 12-27) with 95% confidence interval.....	111
Figure 12: Percentage of participants with high emergency department reliance score (>0.33) by age	111
Figure 13: Mean age at transfer to adult sickle cell clinic.....	112
Figure 14: Continuity of care after transfer to adult care	113

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1. Health Care Transition in Sickle Cell Disease: An Introduction

1.1 Overview of Sickle Cell Disease and Health Care Transition

Sickle cell disease (SCD) is the most common inherited blood disorder in the United States predominantly affecting African Americans with a population estimate of 100,000 (Hassell, 2010). SCD is a complex chronic condition resulting in a variety of complications including vasoocclusive crises, chronic pain, acute chest syndrome, stroke, among others (Miller & Meier, 2012). These complications occur across an unpredictable trajectory which varies among individuals and within the same individual over time (Miller & Meier, 2012) making condition management more challenging.

Over the past three decades, advances in the care for children with SCD have increased their life expectancy from the teens to the late forties (Lanzkron, Carroll, & Haywood, 2013; Quinn, Rogers, McCavit, & Buchanan, 2010). However, concurrent improvement in the quality of life and attention to the unique physiological and psychosocial needs of the surviving adolescents and young adults (AYAs) with SCD remain suboptimal (Christie & Viner, 2009; DiNapoli & Murphy, 2002; Kraaij & Garnefski, 2012; Maslow et al., 2011; Miauton et al., 2003; Suris & Parera, 2005). For example, compared to healthy peers who were the same age, gender, and ethnicity, AYAs with SCD were less likely to graduate from college (Maslow et al., 2011), less

likely to be employed (Abrams , Phillips, & Whitworth, 1994; Bediako , Lavender, & Yasin, 2007; Gil et al., 2004; Maslow et al., 2011), and more likely to be at risk for social isolation (DiNapoli & Murphy, 2002; Olsson et al., 2003) and risky behavior such as drunk driving and drug abuse (Miauton et al., 2003; Suris & Parera, 2005). Compared to younger children with SCD, AYAs had worse health outcomes including higher rates of complications such as vasoocclusive crises and strokes (Darbari et al., 2012; Lanzkron et al., 2013; Miller & Meier, 2012; Platt et al., 1994; C. T. Quinn et al., 2010); higher rates of affective conditions such as depression and anxiety (Benton et al., 2007; Jerrell et al., 2011), with reported rates as high as 45% (Jerrell et al., 2011); higher rate of health care utilization (Brousseau et al., 2010, Fosdal & Wojner-Alexandrov, 2007; Lotstein et al., 2008); and higher risk for mortality (Hassell, 2010; Lanzkron et al., 2013). With more than 95% of children with SCD surviving into adulthood (Lanzkron et al., 2013; Quinn et al., 2010), health care services centered around the goal of improving outcomes in this vulnerable age group of AYAs become essential. Consequently, knowledge development centered upon the needs of AYAs and the health care services they should receive as they transition to adult health care and more autonomous self-management of their condition is needed.

Health care transition can be defined as *“the purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from child-centered to*

adult-oriented health-care systems" (Blum et al., 1993, p.570). The goal of health care transition is to maximize the lifelong functioning and potential of the AYA through the provision of high-quality, developmentally appropriate health care services that continue uninterrupted as the individual moves from adolescence to adulthood (American Academy of Pediatrics, American Academy of Family Physicians, & American College of Physicians-American Society of Internal Medicine, 2002; American Academy of Pediatrics, American Academy of Family Physicians, & American College of Physicians, 2011; Rosen, Blum, Britto, Sawyer, & Siegel, 2003). The process of transition should begin early in adolescence, around age 12 years (American Academy of Pediatrics et al., 2002), with transition readiness assessment and education on self-management, and independent life skills, among other skills (Cooley, 2013; McPheeters et al., 2014; White, McManus, McAllister, & Cooley, 2012), and should continue throughout adolescence culminating in young adulthood with the transfer of care from pediatric providers to adult providers and the integration into adult care.

The need for transition services has been endorsed by many pediatric, adolescent, and adult health organizations (American Academy of Pediatrics et al., 2002; American Academy of Pediatrics et al., 2011; Rosen et al., 2003) and several transition programs are available to prepare AYAs to transition to adult care (McPheeters et al., 2014). Despite availability, the effectiveness of these programs in improving outcomes is

not known (Sharma, O'Hare, Antonelli, & Sawicki, 2014). As a result, several federal agencies in the United States and a variety of professional health organizations have emphasized the need for outcome measures in health care transition that are sensitive to change and can identify individual predictors for successful transition (McPheeters et al., 2014; Rosen et al., 2003). However, few studies have examined health care transition in AYAs with SCD (Andemariam et al., 2014; Jordan et al., 2013; Newland, 2008; Telfair et al., 2004; Telfair et al., 1994; Treadwell et al., 2011) and most studies that examined transition outcomes in AYAs with chronic illnesses focused on patient reported outcomes such as satisfaction with the process (McPheeters et al., 2014) and were cross-sectional descriptive studies. While transfer to adult care has been reported as a measure of successful transition (McPheeters et al., 2014), transfer as an outcome falls short in evaluating successful integration into adult care. A better understating of health care transition in SCD is still needed.

1.2 Shifting Self-management in Preparation for Transition

Developing the capacity for chronic illnesses self-management is an essential competency required of the AYA for successful transition to adult care (Kieckhefer & Trahms, 2000; White et al., 2012). Transition is a twofold process that begins with an evolving shift of skills, responsibility, and accountability of chronic condition management from the parent to the AYA, and then preparing the AYA to transfer to

adult care (White et al., 2012). During childhood, parents assume the primary responsibility of the health management of their child's condition, but with time, parents need to progressively shift that responsibility towards the AYA (Sawyer & Aroni, 2005; White et al., 2012). The shifting from parent management to independent AYA self-management is complex. In addition to adequate knowledge about their condition and the technical self-care skills (such as refilling prescriptions), independent self-management by AYAs requires adequate capacity in problem solving and decision making (Bell & Sawyer, 2010; Sawyer & Aroni, 2005). For AYAs with SCD, assuming independent self-management might be more challenging given the complex and unpredictable nature of SCD.

Very few studies have examined self-management in individuals with SCD (Andemariam et al., 2014; Jordan, Swerdlow, & Coates, 2013; Newland, 2008; Telfair, Ehiri, Loosier, & Baskin, 2004; Telfair, Myers, & Drezner, 1994; Treadwell, Telfair, Gibson, Johnson, & Osunkwo, 2011) and none of these studies examined the shifting of responsibility of SCD management from the parent to the AYA with SCD. Studies that explored self-management strategies for adults with SCD had identified a variety of successful strategies including symptom self-awareness, building collaborations with providers, and use of coping strategies (Jenerette & Phillips, 2006; Tanabe et al., 2010). Jenerette and Phillips (2006) found a difference in the strategies used based on the

individual's age. However, given the dearth of investigations exploring self-management SCD strategies among younger age groups with SCD, the extent of how helpful and how well-used these strategies are remains unclear. AYAs with SCD need to increasingly engage in self-management of their condition to improve their disease, social, psychological, and vocational outcomes (Aujoulat et al., 2014; Sattoe et al., 2015). Developing a deeper understanding of the challenges adolescents with SCD and their parents face in shifting the management responsibility from the parent to the adolescent and the strategies they use in overcoming these challenges can help us understand the facilitators and barriers to more independent self-management in the AYA and can guide the development of interventions to facilitate the shifting of management responsibility.

1.3 Health Care Utilization

While the annual cost of health care for individuals with SCD is estimated to be as high as \$1.1 billion (Kauf, Coates, Huazhi, Mody-Patel, & Hartzema, 2009), the patterns of health care utilization for AYAs with SCD in relation to their health care transition remain poorly understood. The limited body of research that explored health care utilization in AYAs with SCD indicated that AYAs might be at risk for high rates of recurrent hospitalizations and emergency department encounters (Blinder et al., 2015; Brousseau, Owens, Mosso, Panepinto, & Steiner, 2010; Kauf et al., 2009; Wolfson et al.,

2012). One study found that 18 to 30-year-old individuals had the highest hospitalization and re-hospitalization rates as well as the highest emergency department encounters compared to both younger and older individuals with SCD (Brousseau et al., 2010). However, most of these studies were limited to one or two services and did not explore health care utilization across services. As a result, there is a limited understanding of the patterns of health care utilization across services in AYAs with SCD particularly in relation to health care transition.

There is also a limited understanding of the factors that might contribute to increased health care utilization in the population of transitioning AYAs with SCD. Research exploring individual characteristics, such as disease severity and gender, and their association with patterns of health care utilization in AYAs with SCD is inconclusive (Aisiku et al., 2009; Epstein, Yuen, Riggio, Ballas, & Moleski, 2006). Other factors that might be associated with increased utilization and warrant further exploration include contextual factors such as the type of health insurance (Brousseau et al., 2010; Schlenz, Born, Lackland, Adams, & Kanter, 2016) and location of residence (Schlenz et al., 2016; Telfair, Haque, Etienne, Tang, & Strasser, 2003; Wolfson et al., 2012).

In summary, there is limited research that explored health care utilization in AYAs with SCD. In particular, there is a limited understanding of the patterns of health care utilization across services in AYAs with SCD who are transitioning to adult care

and the factors that might be associated with these patterns. Decreasing health care cost for AYAs with SCD requires a better understanding of the patterns of health care utilization in SCD and the factors associated with these patterns.

1.4 Theoretical Framework- Bronfenbrenner's Bioecological Theory of Human Development

Transition to adult care is viewed in a developmental perspective by many pediatric, adolescent, and adult professional health organizations (American Academy of Pediatrics et al., 2002; Rosen et al., 2003). A bioecological perspective such as Bronfenbrenner's bioecological theory of human development provides a useful framework to examine health care transition for AYAs with SCD. Bronfenbrenner's bioecological theory has undergone several revisions (Bronfenbrenner, 2005; Bronfenbrenner & Ceci, 1994; Bronfenbrenner & Morris, 2006; Rosa & Tudge, 2013; Spencer, Dupree, & Hartmann, 1997) and in its latest form has four main interrelated components exerting synergistic influences on human development. These components are process (proximal processes), person, context, and time (Bronfenbrenner, 2005; Bronfenbrenner & Morris, 2006).

Proximal processes (process) are at the core of the theory constituting the major forces driving human development. Proximal processes are reciprocal interactions between the developing individual and the persons, objects, and symbols in their environments, both immediate and distal. To influence development, these interactions

should be regular, progressively more complex, and occur over prolonged periods of time. The direction, form, and content of proximal processes are influenced by the other three main components in the theory: person, context, and time (Bronfenbrenner & Morris, 2006).

Personal characteristics of the developing individual (person) influence proximal processes by either encouraging or discouraging a reaction from the environment. Personal characteristics are categorized into demand characteristics (such as race and gender), resource characteristic (such as social, cognitive, and financial resources), and force characteristics (such as impulsiveness, aggression, and self-efficacy) (Bronfenbrenner, 2005; Bronfenbrenner & Morris, 2006).

The context consists of four interrelated levels of the environment- immediate and distal to the developing individual. First, the microsystem is the most immediate environment with respect to the developing individual, and it consists of the “pattern of activities, roles, and interpersonal relations experienced by the developing person” (Bronfenbrenner, 2005, p.148) during face to face interaction with other individuals. Second, the mesosystem is the linkage processes that are taking place between two microsystems, or two settings, containing the developing individual such as the home and the health care setting. Third, the exosystem is the linkages and process between two settings, one of which does not contain the developing individual. The exosystem

exerts indirect developmental influences on the individual through the influences it has on others in the microsystem. Fourth, the macrosystem is the overarching characteristics of the culture or other social context where individuals share a common belief or value system -the “societal blueprint” of the social context such as economic, social, legal and political systems (Bronfenbrenner & Ceci, 1994). The developing individual is in the innermost circle of this nested structure of environmental systems. These systems are interconnected and the degree of influence on the developing individual is not proportionate to their level of proximity (Bronfenbrenner & Morris, 2006).

The concept of time is broad (Rosa & Tudge, 2013) and temporal influences exist at the micro-, meso-, and macro- levels (Bronfenbrenner & Morris, 2006). Bronfenbrenner adopted Glen Elder’s principles of the influence of time on human development. Specifically, timing in terms of when the events occur in a person’s life is a major influencing factor on human development (Bronfenbrenner & Morris, 2006). Time influences can be either disruptive or provide an opportunity for the developing individual to develop and grow.

Bronfenbrenner’s bioecological theory of human development with its focus on development processes within the context and across time provides a useful framework for understanding the complex multilevel phenomenon of health care transition. First, health care transition depends on such reciprocal interactions within the AYA’s

environment (process). Health care transition is shaped by the interactions that the AYA with SCD has with family members and health care providers to build the skills needed for independence (Porter, Graff, Lopez, & Hankins, 2014). AYAs who are transitioning to adult care are moving from a family-centered model in pediatric care, where parents assume a lead role in care, to a patient-centered model in adult care, where the AYA is expected to be the primary person responsible for care (White et al., 2012). For the AYA this entails acquiring the skills of self-management as well as the skills of navigating the more complex adult health care system. For the parent, this entails progressively releasing the responsibility of care to the AYA (Kieckhefer & Trahms, 2000; White et al., 2012) and for the health system this entails facilitating the shifting of responsibility for care from the parent to the AYA. Family and health care provider interactions with the AYA have the potential of supporting the adolescent's transition to adult care by fostering their progressive independence in self-management and navigation of the health care system (Bronfenbrenner & Morris, 2006) and addressing the challenges that might arise. Furthermore, transition should not be an abrupt event. It should be a process evolving over time (de Montalembert & Guitton, 2014; Hauser & Dorn, 1999; Treadwell et al., 2011; White, 1999) a process of skill building in self-management and independence nurtured and supported by the AYA's family, health care system, and community.

In summary, a bioecological developmental approach focusing on the progressively more complex interactions with key individuals, such as family and friends, and systems, such as the health care system, within different and multilevel contexts in the AYA's environment are critical to a comprehensive understanding of a complex and dynamic developmental process such as health care transition.

1.5 Purpose Statement and Research Aims

The purpose of this dissertation was to provide a better understanding of health care transitions in AYAs with SCD. This purpose was achieved through an examination of the state of the science on health care transition in AYAs with SCD, the challenges of shifting self-management from the parent to the adolescents with SCD prior to transition to adult care, and of health care utilization during transition. Knowledge gained from this dissertation will enhance the understanding of the development of independent self-management in adolescents with SCD and their health utilization in relation to health care transition. These findings will be critical to guide the development of interventions that enhance the capacity of adolescents with SCD to independently manage their condition and successfully transition into adult-based care. The purpose of the dissertation was achieved through the following aims:

1.5.1 Aim one

Introduce the problem and significance of independent self-management and transition to adult care in AYAs with SCD (Chapter one).

1.5.2 Aim two

Conduct an integrative review to summarize and synthesize the current state of the science on health care transition in AYAs with SCD (Chapter two).

1.5.3 Aim three

Explore and describe the challenges that adolescents (12-18 years old) with SCD and their parents have and the strategies they use to shift the management of SCD from the parent to the adolescent prior to transition to adult care using a descriptive qualitative study design (Chapter three). Specific research questions were:

1. What are the adolescents' challenges with shifting the management of SCD?
2. What are the adolescents' strategies with shifting the management of SCD?
3. What are the parents' challenges with shifting the management of SCD?
4. What are the parents' strategies with shifting the management of SCD?

1.5.4 Aim four

Identify and describe trajectories of health care utilization of AYAs (12 to 27 years old) with SCD and examine the association between trajectory groups and individual characteristics, contextual factors, and transfer related factors. success. The

overall goal was to identify and define successful transition for AYAs with SCD. This was achieved through a longitudinal databased analysis examining AYAs with SCD who received care at Duke Health (Chapter 4). Specific research aims were:

- 1 Identify and describe the trajectories of health care utilization of transitioning AYAs (12 to 27 years old) with SCD, specifically:
 - 1.1 What are the classes of AYAs with SCD who had similar health care utilization trajectories during transition to adult care?
 - 1.2 How does transfer to adult care occur?
- 2 Examine the association between the utilization trajectory groups and the individual characteristics, contextual factors, and transfer related factors.
Specifically:
 - 2.1 What are the influences individual characteristics, contextual factors, and transfer related factors on the probability of class membership for health care utilization?

1.5.5 Aim five

Synthesize the findings from each of the chapters in this dissertation and present implications and recommendations to enhance the capacity of self-management in AYAs with SCD and the factors that facilitate or hinder their successful transition into adult-based care in relation to health care utilization (Chapter 5).

2. Health Care Transition for Adolescents and Young Adults with Sickle Cell Disease: State of the Science Integrative Review

Health care transition, defined as *“the purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from child-centered to adult-oriented health-care systems”* (Blum et al., 1993, p.570), aims to maximize the lifelong functioning and potential of the adolescents and young adults (AYAs) with chronic conditions through the provision of high-quality, developmentally appropriate health care services that continue uninterrupted as the individual moves from adolescence to adulthood (American Academy of Pediatrics, et al., 2002; American Academy of Pediatrics, et al., 2011; Rosen, et al., 2003). Health care transition is a relatively new field of research and practice (Betz et al., 2014) and the science on what constitutes successful transition, what factors facilitate or hinder successful transition, and the impact that transition has on the AYA’s outcomes is still emerging (McPheeters et al., 2014).

Due to recent advances in the care of children with sickle cell disease (SCD), the vast majority of these children are expected to survive into adulthood (Lanzkron, Carroll, & Haywood, 2013; Quinn, Rogers, McCavit, & Buchanan, 2010) and to transfer from pediatric to adult care at some point during late adolescence or early adulthood. Very few reviews have been published examining health care transition in AYAs with SCD and these reviews were either limited in scope or in methodology (Baskin et al., 1998; Crosby, Quinn, & Kalinyak, 2015; Jordan, Swerdlow, & Coates, 2013; Molter & Abrahamson, 2014; Yacobovich & Tamary, 2014). For example, a recent review by Jordan and colleagues (2013) only included research identified through one database (PubMed) and was limited to research published over ten years (2001-

2011). Another review by Molter and Abrahamson (2014) focused only on self-efficacy, transition, and SCD outcomes, while few other reviews included SCD among other chronic conditions and did not specifically focus on SCD (Betz, Lobo, Nehring, & Bui, 2013; Betz, O'Kane, Nehring, & Lobo, 2016; Bryant, 2009; Callahan, Feinstein, & Keenan, 2001; Chu, Maslow, von Isenburg, & Chung, 2015; Leader & Raanani, 2014; Nehring, Betz, & Lobo, 2015; Perry, 2014; Prior, McManus, White, & Davidson, 2014; Rachas et al., 2016; Schwartz et al., 2014; Waldboth, Patch, Mahrer-Imhof, & Metcalfe, 2016; Zhou, Roberts, Dhaliwal, & Della, 2016). A better understanding of the current state of the science on health care transition in SCD is needed to guide future research. Therefore, the purpose of this integrative review was to provide a comprehensive review of current state of the science on health care transition for AYAs with SCD.

2.1 Theoretical Framework

The health care transition (HCT) model (Betz et al., 2014) was recently developed as a guiding framework for transition research and practice. The model takes a non-categorical approach to describe variables, health care transition processes, mediators and moderators that might influence the outcome of health care transition across conditions. The desired outcomes are acquiring the developmental competencies necessary for transition to adulthood and effective integration in adult health care. These outcomes are achieved through the interaction of four domains: the individual domain, the family/social support domain, the environmental domain, and the health care system domain (Betz et al., 2014).

The individual domain includes characteristics of the individual that might influence the individual's ability to attain the knowledge and skills necessary for successful transition. These

characteristics include demographic characteristics, condition stability and the natural course of the disease process, development competencies, personality processes, and self-management capacities. The family/ social support domain includes the level of family support and the level of social support. The level of family support includes the parent-child relationship, family resources, family culture, family health status, family competency, and family involvement in care. Social support include the characteristics and availability of social support networks and the availability and access to the social environment. The environment domain includes systems, services, and resources made available by the community and the educational system serving the individual. Ability to access these community and educational resources will influence the individual's health care transition outcomes. Finally, the health Care system domain includes the pediatric health care system, the adult health care system, access to adult health care insurance plan, patient-provider relationship, and health care payment/ reimbursement structures and models (Betz et al., 2014).

2.2 Methods

A comprehensive literature search was conducted in PubMed, Comprehensive Index to Nursing and Allied Health Literature (CINAHL), psycINFO, Scopus, and Web of Science to identify relevant literature on health care transition in AYAs with SCD. With the help of a medical librarian, we identified and used a combination of sickle cell disease, adolescent, young adult, and health care transition search terms. The exact search terms used for each of the databases and the number of articles each search yielded are presented in table 1. The searches were limited to the English Language and peer reviewed journals (when the peer review limit was an option in the database).

Table 1: Database search terms and results

Database	Search terms	Articles
PubMed	(Adolescent[mh] OR youth[tiab] OR adolescent[tiab] OR adolescents[tiab] OR adolescence[tiab] OR teen[tiab] OR teens[tiab] OR teenage[tiab] OR teenager[tiab] OR teenagers[tiab] OR young adult[mh] OR "young adult"[TiAb] OR "young adults"[TiAb] OR "emerging adult"[TiAb] OR "emerging adult"[TiAb]OR "young people"[tiab]) AND ("anemia, sickle cell"[MeSH] OR "sickle cell"[tiab]) AND ("Continuity of Patient Care"[mh:noexp] OR "Transition to Adult Care"[mh] OR transition[tiab] OR transitions[tiab] OR transitioning[tiab] OR transitional[tiab]) <i>Limited to English and human</i>	79
CINAHL	((MH "Adolescence+" OR MH "Young Adult" OR TI (youth OR adolescent OR adolescents OR adolescence OR teen OR teens OR teenage OR teenager OR teenagers OR "young adult" OR "young adults" OR "emerging adult" OR "emerging adult" OR "young people")) OR AB (youth OR adolescent OR adolescents OR adolescence OR teen OR teens OR teenage OR teenager OR teenagers OR "young adult" OR "young adults" OR "emerging adult" OR "emerging adult" OR "young people")) AND(MH "Anemia, Sickle Cell" OR TI ("sickle cell") OR AB ("sickle cell")) AND ((MH "Continuity of Patient Care") OR (MH "Health Transition") OR TI(transition OR transitions OR transitioning OR transitional) OR AB (transition OR transitions OR transitioning OR transitional)) <i>Limited to English and peer reviewed</i>	55
PSYINFO	((TI (youth OR adolescent OR adolescents OR adolescence OR teen OR teens OR teenage OR teenager OR teenagers OR "young adult" OR "young adults" OR "emerging adult" OR "emerging adult" OR "young people")) OR AB (youth OR adolescent OR adolescents OR adolescence OR teen OR teens OR teenage OR teenager OR teenagers OR "young adult" OR "young adults" OR "emerging adult" OR "emerging adult" OR "young people")) AND(DE "Sickle Cell Disease" OR TI ("sickle cell") OR AB ("sickle cell")) AND (DE "Continuum of Care" OR TI(transition OR transitions OR transitioning OR transitional) OR AB (transition OR transitions OR transitioning OR transitional)) <i>Limited to English and peer reviewed</i>	30
SCOPUS	(TITLE-ABS-KEY (youth OR adolescent OR adolescent OR adolescence OR teen OR teens OR teenage OR teenagers OR teenagers OR "young adult" OR "young adults" OR "emerging adult" OR "emerging adult" OR "young people") AND TITLE-ABS-KEY ("sickle cell") AND TITLE-ABS-KEY (transition OR transitions OR transitioning OR transitional)) <i>Limited to English, article, review, and article in press</i>	91
Web of Science	(youth OR adolescent OR adolescent OR adolescence OR teen OR teens OR teenage OR teenagers OR teenagers OR "young adult" OR "young adults" OR "emerging adult" OR "emerging adult" OR "young people") AND ("sickle cell") AND (transition OR transitions OR transitioning OR transitional) <i>Limited to English</i> <i>Excluded meeting abstracts, editorial material, letter, and conference proceedings</i>	109

The search yielded 79 articles in PubMed, 55 articles from CINAHL, 30 articles in PsycINFO, 91 in Scopus, and 109 in Web of Science. The results of the searches were merged and duplicates were removed, which resulted in a total of 160 unique articles. Then, titles and abstracts were carefully screened for relevance to health care transition in AYAs with SCD. Articles were excluded if they were not empirical research, not peer reviewed, or did not address health care transition in AYAs with SCD. The full text of 55 articles was reviewed. Further articles on health care transition in AYAs with chronic illness were excluded if the sample included AYAs with SCD but findings on AYAs with SCD were not presented separately (n=16). A final set of 39 articles was included in this integrative review (figure 1).

The matrix method (Garrard, 2011) was used to examine the literature for common themes across studies and identify the relation and difference between them. The 39 articles were abstracted into a matrix with 17 headings: Author, year, purposes, hypothesis, design, sample, inclusion criteria, exclusion criteria, definition of concepts/key terms, study framework, data collection/instruments, reliability/validity, findings, strengths, weaknesses, suggested areas for new research, and significance to AYAs with SCD.

The articles were reviewed systematically and thoroughly, and the content was analyzed using the mixed research synthesis (Sandelowski, Voils, & Barroso, 2006) approach to integrate both qualitative, quantitative, and mixed method studies on health care transition of AYAs with SCD. Using this approach allowed us to integrate research findings regardless of study methodology and allowed for a broader understanding of health care transition in AYAs with SCD. Specifically, we used the integrated design where we transformed findings from

quantitative studies to qualitative finding (Sandelowski et al., 2006). A deductive approach was then used to organize the literature by themes of the HCT model (Betz et al., 2014).

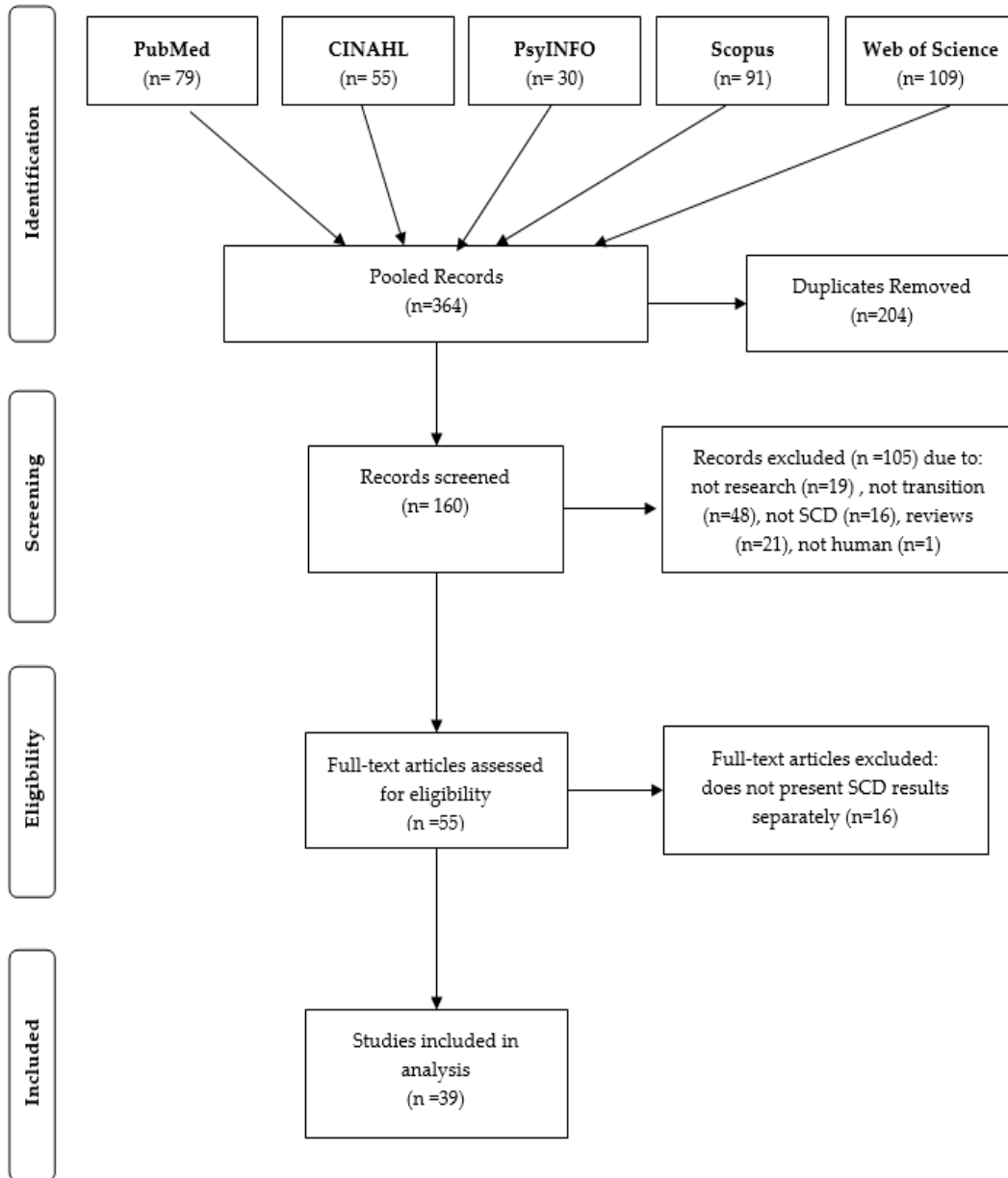


Figure 1: Prisma flow diagram

2.3 Results

2.3.1 Description of studies

The majority of health transition research in AYAs with SCD involved the individual and health care system domains. Few studies examined health care transition outcomes or the family/social support domains, and we could not locate any studies that examined the environmental domain. Almost all the studies were conducted in the United States, except for one study which was jointly conducted between the United States and the United Kingdom. Studies used either qualitative or quantitative designs, and only one study used mixed methods. Table 2 summarizes the studies designs, number of participants, and main findings.

2.3.2 Health care transition outcomes

Very few studies examined transition outcomes as outlined in the HCT model. Achieving independence was the only developmental competency studied and only two studies included independence- one as an outcome and the other as a factor contributing to the AYA's attendance at the adult clinic. Similarly, integration into adult care was examined in only three studies.

Overall, there is not enough evidence on what factors contribute to the achieving of independence or whether independence is associated with keeping adult appointments in the AYA with SCD. Newland and colleagues (2008) examined the relation between knowledge about SCD, severity of disease, and family relationships to the level of independence in adolescents with SCD. They defined independence in terms of high autonomy scores and found that, while the majority of their adolescents had high autonomy scores, the correlation between family relationships, severity of disease index, knowledge of SCD and the adolescent's

autonomy scored accounted for only 25% of the variance for independence. On the other hand, Wojciechowski and colleagues (2002) examined whether achieving or not achieving independence in the AYA with SCD influenced their rate of attendance at the adult clinic. They defined independence as the ability of the AYA to have health care coverage, enroll or pursue higher education or have a vocation, and having self-efficacy for managing care. Although they reported that all their AYAs had some form of health care coverage, and were enrolled as students or had a vocation, none of these factors were associated with keeping adult appointments. However, findings from both studies should be interpreted with caution. Both studies had small sample sizes and the study by Wojciechowski and colleagues (2002) only recruited participants who were not lost to follow-up. As a result, more research is need to understand how AYAs with SCD achieve adult competencies and what are the factors that facilitate or hinder this process.

Evidence on the second outcome in the model is also inconclusive. The three studies that examined integration into adult care evaluated the effectiveness of transition programs in integrating the AYAs with SCD into adult care. In two studies integration into adult care was examined in terms of whether the AYAs kept their first adult appointment and the duration of time between the last pediatric and first adult appointment (Hankins et al., 2012; Wojciechowski et al., 2002), while the third study did not provide any operational definition for “continuity in adult care” (Andemariam et al., 2014). The length of time between the last pediatric and the first adult appointment was widely variable between participants- ranging between one and 29 months (Wojciechowski et al., 2002) and one day to 10 months (Hankins et al., 2012). While Wojciechowski and colleagues (2002) found no difference between those who completed a

transition program and those who did not in terms of keeping their first adult appointment, Hankins and colleagues (2012) found that 74% of those who participated in their transition program versus 33% of those who did not participate had their first visit to the adult care provider within three months of their last pediatric appointment. Similarly, Andemariam and colleagues (2014) reported that 68% of their sample established continuity in adult care after completing a transition program- although it is not clear how continuity of care was defined. Furthermore, participants who kept more adult appointments after transfer had higher self-efficacy scores, indicating that higher self-efficacy might be associated with better integration in adult care (Wojciechowski et al., 2002). However, all of these studies had small sample sizes and as a result provided weak evidence on the effectiveness of transition programs in improving integration into adult care.

In summary, very few studies examined health care transition outcomes as defined by the HCT model. These studies had methodological issues in terms of sample size and/ or the sample inclusion criteria making it difficult to draw conclusions from findings.

2.3.3 Individual domain

The individual domain was one of the most studied domains among the 39 articles included in this review. The range of individual factors studied included: demographic characteristics, disease complexity and course, developmental competencies, personality processes, and self-management and were usually examined from the perspective of barriers and facilitators to transition or transition readiness.

Demographic characteristics. The association between transition on one hand and age, gender, employment, education, socioeconomic status, and distance from the clinic on the other hand were examined in several studies.

Age. Almost all studies that examined age found a positive association between age and health care transition (Andemariam et al., 2014; McPherson, Thaniel, & Minniti, 2009; Speller-Brown et al., 2015; Treadwell et al., 2016). Specifically, older age was associated with increased transition readiness (McPherson et al., 2009; Speller-Brown et al., 2015; Treadwell et al., 2016) and perceived readiness to assume more responsibility for care by both AYAs and their parents (Speller-Brown et al., 2015). Further, two studies found that AYAs knowledge about their disease increased with age (Abel et al., 2015; Griffin et al., 2013). However, later age at the start of the transition process was found to be negatively associated with transition success. Specifically, starting the transition process after age 21 was associated with lower chances of successful transition (Andemariam et al., 2014).

Gender. Research on the association between gender and transition is inconclusive. While McPherson and colleagues (2009) found that gender was associated with mean transition readiness scores, with females reporting higher levels of anticipated difficulties with perceived transition readiness than males, Andemariam and colleagues (2014) did not find any association between gender and transition success. Other studies also found no significant effect of gender on educational needs (Abel et al., 2012), transition knowledge, or autonomy scores (Newland et al., 2008).

Race, distance to clinic, and insurance. The association between race, distance to clinic, and type of insurance and transition was examined in only one study. Andemariam et al. (2014)

examined transition success following the implementation of a combined transition program and found that demographic factors including race and type of insurance were not associated with transition success. However, they found that distance to the clinic was negatively associated with transition success. Specifically, living more than 20 miles were associated with lower chances of successful transition (Andemariam et al., 2014).

Socioeconomic status. Three studies examined the effect of socioeconomic status on transition from the perspective of AYAs with SCD and health care providers. In one study, around 40% of young adults with SCD reported that they were unemployed and another 20% had a disability and were unemployed (Mennito, Hletko, Ebeling, Amann, & Roberts, 2014). These young adults were concerned about keeping a job, paying for medical care, and arranging for transportation to appointments should they transition to adult care (Mennito et al., 2014). The two other studies examined the perspectives of health care providers (HCPs) and found that HCPs believed that adequate housing, financial stability, and transportation might influenced transition readiness in AYAs with SCD (Mulchan, Valenzuela, Crosby, & Diaz Pow Sang, 2016; Stollon et al., 2015). Annual income and education level were not significantly associated with readiness to assume responsibility, parent involvement in care, and overall readiness to transfer from pediatric to adult care in one study (Speller-Brown et al., 2015).

Disease complexity and course. The impact of disease severity on transition readiness was examined in few studies (Abel et al., 2015; Andemariam et al., 2014; McPherson et al., 2009; Mulchan et al., 2016; Stollon et al., 2015). McPherson and colleagues (2009) found that low mean transition readiness score was associated with increased disease severity while Andemariam and colleagues (2014) found an association between genotype and transition success.

Specifically, AYAs with the SC or S beta zero genotype had lower rates of transition success. While the lack of a need for blood transfusions was associated with unsuccessful transition, utilization of hydroxyurea, frequency of vasoocclusive crisis and acute chest syndromes episodes were not associated with transition success in one study (Andemariam et al., 2014).

Other studies reported on the perspectives of AYAs and/ or HCPs. HCPs believed that challenges related to the progression of disease and the occurrence of disease complications affected transition, particularly in the presence of cognitive deficits associated with cerebral infarcts (Mulchan et al., 2016; Stollon et al., 2015) and the impact cognitive deficits might have on the AYA's ability to understand the disease process and navigate the health care system (Stollon et al., 2015). Further, Abel et al. (2015) found that the presence of cerebrovascular injury had a significant effect on the percentage of AYAs who reported that they need more practice on areas for transition.

Finally, one national study examined mortality rates in individuals with SCD and found that while mortality rates significantly decreased in children less than 19 years, mortality rates increased for young adults with SCD after age 19, indicating a higher risk for mortality during transition (Hamideh & Alvarez, 2013).

Developmental competencies. Developmental competencies, and more specifically, psychosocial level of function was reported in two studies as an important factor for successful transition (Mulchan et al., 2016; Stollon et al., 2015). Both studies reported on the perspectives of HCPs who believed that maturity was significant facilitator for successful transition. Interestingly, Mulchan and colleagues (2016) found that most adolescents in their study felt that they would be ready to transition at age 18, while most providers expressed that self-

management tasks should be tailored to the AYA's developmental level so they can gradually acquire skills necessary for managing SCD and transition to adult care (Mulchan et al., 2016). HCPs believed that developmental milestones might interfere with the AYA's transition readiness. Specifically, developmental milestones such as attending college, obtaining employment, and starting romantic relationships might compete with health care transition making the transition process more challenging (Mulchan et al., 2016).

Personality processes. Personality processes reported in the studies reviewed included personal attributes and self-efficacy. Only one study examined personal attributes associated with successful transition and it was examined from the HCP perspective. In this study, HCPs believed that AYAs with positive personal attributes such as proactivity, goal-orientation, and adaptive emotional functioning (adequate coping, absence of psychopathology, and motivation) would be more successful at transition (Mulchan et al., 2016).

Self-efficacy was reported in four studies. Self-efficacy was found to be positively associated with readiness to transition (Treadwell et al., 2016) and keeping more adult appointments after transfer (Wojciechowski et al., 2002). Anie and Telfair (2005) explored factors associated with higher self-efficacy in two samples of adolescents with SCD in the United States and the United Kingdom. They found no association between self-efficacy and demographic factors such as age and education level. However, self-efficacy was associated with physical symptoms indicating that disease severity might influence self-efficacy (Anie & Telfair, 2005).

Self-management. Few studies explored self-management in AYAs with SCD in relation to health care transition. These studies explored assumption of responsibility by the AYA, disease knowledge, and adherence.

Assumption of responsibility. Assumption of responsibility of care was in few studies from the perspective of the AYA, parents, and HCPs.

AYAs, parents, and health care providers acknowledged the need of the AYA's to assume more responsibility of their care before transition (Kayle, Tanabe, Shah, Baker-Ward, & Docherty, 2016; Mulchan et al., 2016; Porter, Graff, Lopez, & Hankins, 2014; Speller-Brown et al., 2015), including pain management skills and health care navigation skills such as obtaining health insurance coverage and scheduling appointments (Mulchan et al., 2016). AYAs also expressed the desire to have more opportunities to practice their independence, such as attending clinic appointments (Porter et al., 2014). However, Speller-Brown et al. (2015) found that AYAs perceived that their parents were more responsible than they were for their care, and although they took responsibility sometimes for some but not all management, they often relied on parents who were very involved in the health care management of the AYA's condition. AYAs also rated themselves to be "somewhat to "not ready" at all to assume responsibility of care while parents believed their AYAs were "somewhat" to "often responsible" for their care. The perceptions of AYAs and parents that the AYAs can assume responsibility of care increased with the AYA's age. Speller-Brown et al. (2015) also found that increased readiness for transition was associated with the amount of responsibility the AYAs assumed for their care.

Disease knowledge. Adequate SCD and medical history knowledge was emphasized as an important requirement for successful transition by both AYAs and their HCPs (Mulchan et

al., 2016). Transition preparation programs were successful in improving the AYA's knowledge about SCD and their medical history (Griffin et al., 2013; Zhao et al., 2016). However, programs should be tailored to the needs of the AYA (Zhao et al., 2016). Studies that assessed knowledge found that AYAs were knowledgeable on most but not all aspects of their condition or medical history (Sobota et al., 2014; Zhao et al., 2016). Areas needing additional education included: ethnicities affected by SCD (Smith, Lewis, Whitworth, Gold, & Thornburg, 2011), inheritance of SCD (Newland, 2008; Smith et al., 2011), treatment options (Newland, 2008), and independent living and health benefits skills sets (Sobota et al., 2014). AYAs with cerebrovascular disease had these areas included in their learning needs: living arrangements, money management, vocational skills, and health care skills (Abel et al., 2015).

Adherence. Only one study explored adherence to medical management. Wojciechowski et al. (2002) found no significant difference between participants who received a transition program versus those who did not on self-reported compliance with management.

Overall, there is evidence that indicates that with increasing age, AYAs report improved transition readiness, yet delaying transition beyond age 21 might be problematic to transition success. Findings are inconclusive with regards to the effect of other demographic factors including gender, socioeconomic status, insurance type, race and ethnicity, educational level, and employment on health care transition in AYAs with SCD.

2.3.4 Family/Social support domain

Most of the studies focused on family support rather than the social support domain and included parental, AYA's, and HCP's perspectives.

Parents of AYAs were found to be heavily involved in the care of the AYA (Kayle et al., 2016; Porter et al., 2014; Speller-Brown et al., 2015). Parents felt challenged to decrease their involvement in care to allow the adolescent with SCD the chance to assume more care responsibility (Kayle et al., 2016; Porter et al., 2014) because parents (Kayle et al., 2016; Porter et al., 2014) and siblings (Porter et al., 2014) were concerned about the ability of the adolescent to independently take care of themselves. It appears that parental involvement in care continues to be high throughout adolescence and into young adulthood (Speller-Brown et al., 2015), at least until parents feel that the AYA is old enough and can assume more responsibility of their care (Speller-Brown et al., 2015). Parents' perceptions of their involvement in AYA healthcare behaviors and their perception of AYA overall readiness to assume complete responsibility had strong positive correlations with parents' perceptions of AYA overall readiness to transfer from pediatric to adult care as well (Speller-Brown et al., 2015). Although AYAs were aware of their parental heavy involvement in care (Kayle et al., 2016; Porter et al., 2014; Speller-Brown et al., 2015), AYAs did not perceive parental involvement as an indicator of their assumption of more care responsibility or transition readiness (Speller-Brown et al., 2015) and generally perceived parental involvement as positive and supportive (Kayle et al., 2016; Labore, Mawn, Dixon, & Andemariam, 2017; Porter et al., 2014). Only in one study did AYAs with SCD discuss how SCD interrupted their family, work, and social roles (Matthie, Hamilton, Wells, & Jenerette, 2016).

Family support was believed to be an important facilitator for transition by HCPs as well (Mulchan et al., 2016). HCPs believed a challenging home life and parent education level influenced transition success. On the other hand, having good social support networks and

supportive families were viewed as facilitators for successful transition, sometimes despite the lack of family or social support. Social and family support, while emphasizing the AYA's independent and self-management, were viewed as facilitators of transition (Stollon et al., 2015).

Overall, parents were heavily involved in the AYA's health care. Although this involvement was viewed as supportive by the AYA as well HCPs, it's not clear how parental involvement and the parent-AYA relationship can be utilized to facilitate health care transition for the AYAs with SCD. More research is also needed to understand the role that social support outside of the family structure plays in the AYA's health care transition.

2.3.5 Health care system domain

The health care system domain was among the most studied domains in this review. Areas studied included the AYA-provider relationship, provider technical capabilities related to caring for the AYA with SCD, and availability of health care transition services.

AYA-provider relationship. Most of the studies focused on the AYA-provider relationship, particularly with adult providers, and the concerns that AYAs and their caregivers have with health care transition. There was congruence among AYAs, their caregivers, and HCPs on the importance of a good and supportive AYA-provider relationship for successful transition (Mulchan et al., 2016; Rouse, 2011; Sobota, Shah, & Mack, 2016; Stollon et al., 2015). Studies discussed several barriers and facilitators for a good and trusting AYA-provider relationship and these varied between pediatric and adult providers. Pediatric providers believed that good communication between AYAs and adult providers allowed AYAs to be more comfortable discussing treatment plans and asking questions openly and was a facilitator for transition. Further, pediatric HCPs believed that AYAs with SCD face unique challenges

with building supportive relationships with adult providers and due to stigma and lack of community awareness about SCD. They also believed that AYAs contribute to the lack of community awareness of SCD by their desire for normality and resulting lack of desire to discuss SCD and how it impacts them (Mulchan et al., 2016). On the other hand, adult providers empathized the need for social workers to help AYAs achieve success in the adult system (Stollon et al., 2015) and discussed a trusting relationship between AYAs and HCPs less than pediatric providers (Mulchan et al., 2016). Some adult providers felt that pediatric providers fostered dependency with their relationships with their patients (Hauser & Dorn, 1999; Mulchan et al., 2016).

Several studies assessed AYAs' concerns with health care transition. These concerns included feeling unprepared (Telfair, Ehiri, Loosier, & Baskin, 2004; Tuchman, Slap, & Britto, 2008), fear of leaving a familiar pediatric provider (Hauser & Dorn, 1999; Telfair et al., 2004), fear that the adult provider might not understand their needs (Telfair et al., 2004; Telfair, Myers, & Drezner, 1994), concerns about re-establishing relationships with a new adult team (Latzman et al., 2010; Smith et al., 2011; Tuchman et al., 2008), concerns about the adult care providers' lack of SCD knowledge (Porter et al., 2014), and concerns about using the adult hospital and emergency department (Latzman et al., 2010). AYAs reported feeling scared, fearful, sad, and worried about transition (Porter et al., 2014). Two studies reported on parents' and caregivers' concerns. Parents and caregivers were concerned about leaving behind their previous doctor and about adolescent assuming responsibility for their own care (Hauser & Dorn, 1999; Latzman et al., 2010).

Health care transition services. Research indicated that AYAs with SCD do not receive adequate preparation for transition (Mennito et al., 2014; Williams et al., 2015; Wojciechowski et al., 2002). In a national survey of 30 sickle cell centers in the United States, Sobota and colleagues (2011) found that the majority of centers surveyed had a young transition program and that the specific transition practices varied by center.

Other studies examined the AYA's needs for health care transition. AYAs and their caregivers discussed several recommendations for their HCPs that would assist them in their transition process. These included earlier discussions about transition, meeting the adult team and visiting the adult clinic before transfer, and meeting other people with SCD (Telfair et al., 2004; Tuchman et al., 2008). AYAs and their caregivers emphasized that transition programs should focus on provider support, information about adult care programs, ways to meet adult providers and ways to help adult providers understand the AYA's needs (Telfair et al., 2004). They also expressed a desire to practice being independent (Kayle et al., 2016; Tuchman et al., 2008).

Access to care. AYAs reported that SCD symptoms were complex and taxing requiring various health care service expertise (While & Mullen, 2004). However, they reported difficulties in accessing needed health care after transition (Matthie et al., 2016). The challenge for these AYAs was maintaining their health insurance and changing their health care provider once they have transitioned, which they felt disrupted their quality and continuity of care. They felt that receiving appropriate pain management outside of the sickle cell clinic was challenging because HCPs tended to stigmatize them with drug seeking behaviors. As a result, they

attempted to rely on their self-management for SCD, controlling and preventing pain, and delayed seeking care at hospital or emergency department (Matthie et al., 2016).

Provider technical capabilities. In a large sample of general internist, Okumura et al. (2008) assessed the level of comfort of adult providers with providing care for AYAs with SCD. Less than half of general internists felt that their specialty should take primary care responsibility for adult patients with SCD.

Overall, there is availability of health care transition services for AYAs with SCD, but the practices of these programs tend to vary. AYAs felt they were not appropriately prepared to transition to adult care and had several concerns about the process. They also expressed needs that should be addressed by transition programs to make their transition process easier. The AYA-provider relationship in adult care was considered essential for successful transition.

2.3.6 Additional findings

Transition success. Although not specifically stated as such in the HCT, we think transition success is an implicit outcome in the HCT model. One study defined transition success. Stollon et al. (2015) interviewed 13 providers (physicians, social works, psychologists, and nurses) and found that these providers evaluated successful transition based on health care utilization, quality of life, and continuity on a stable disease trajectory. Successful transition in terms of health care utilization meant that participants made and kept their medical appointments, adhered to their medication regimen, and avoided preventable hospitalizations. Quality of life was described in terms of the AYAs achieving their developmental milestones, such as obtaining postsecondary education, employment, getting married, and having children.

A continuous stable disease trajectory meant that disease severity was comparable or improved after pediatric care.

Health care utilization. Although not a component in the HCT model, health care utilization is an important indicator of health care transition. How AYAs utilize health care services has major implications on their outcomes, including but not limited to integration in adult care.

Few studies examined health care utilization during transition to adult care and they indicate that AYAs who are transitioning to adult care might at risk of higher acute care utilization, tending to utilize emergency department and hospital care instead of ambulatory care for their health care needs (Blinder et al., 2015; Dickerson, Klima, Rhodes, & O'Brien, 2012; Panepinto et al., 2012; Schlenz, Boan, Lackland, Adams, & Kanter, 2016). Schlenz et al. (2016) examined the acute care utilization (hospital and emergency department) of individuals with SCD in the state of South Carolina. They found that young adults had the highest acute care encounters, particularly emergency department encounters, compared to children, adolescents, and older adults. Similarly, Blinder et al. (2015) examined age-related Emergency department visits in a five state Medicaid programs and found that that beginning at age 15 years, emergency department reliance score rose from 0.17 to reach 0.29 at age 22 years, and remained high throughout adulthood, indicating that patients with SCD may rely more heavily on the emergency department for SCD care after they transitioned from pediatric to adult care. Finally, Dickerson and colleagues (2012) found that young adults with SCD had higher hospitalization rates and incurred more costs compared to adolescents. Compared to adolescents, young adults also tended to use multiple hospitals for their care (Panepinto et al., 2012).

2.4 Discussion

The purpose of this integrative review was to provide a comprehensive review of current state of the science on health care transition for AYAs with SCD. We used the health care transition (HCT) model (Betz et al., 2014) and found the model to be a useful to organize findings from our review.

The majority of the research reviewed could be classified as either quantitative or qualitative. Only one study used mixed methods. Most of the research included in this review was conducted in the United States except for one study, which was conducted in the United States and the United Kingdom. We identified few studies that examined outcomes or the family/social support domain and no studies that addressed the environmental domain. Most studies included in this review addressed the individual and/or health care system domains. We also identified transition success and health care utilization as additional domains that were not clearly addressed in the HCT model.

Overall, very few studies examined health care transition outcomes as defined by the HCT model. Independence was the only adult competency studied and all the studies had methodological issues in terms of sample size and the sample inclusion criteria making it difficult to draw conclusions from findings. Integration into adult care was also studied as an outcome of the HCT model, however, the findings are inconclusive. More research is needed in the area of health care transition outcomes.

Table 2: Summary of articles and findings

Authors (year)	Title	Study Design	Sample size	Model Domain	Main findings
Schlenz et al. (2016)	Needs Assessment for Patients with Sickle Cell Disease in South Carolina, 2012	Quantitative	2313	Outcome	Young adults had the highest rates of acute care utilization and 30-day readmissions over 12 months period
Wojciecho-wski et al. (2002)	A Natural History Study of Adolescents and Young Adults With Sickle Cell Disease as They Transfer to Adult Care: A Need for Case Management Services	Mixed	18 AYAs; & their providers	Outcome/individual/Health care system	AYA receive little preparation before transfer; age and pregnancy rather than readiness trigger transfer; follow up ceases after transfer; patients with high self-efficacy maintain adult appointments
Newland et al. (2008)	Factors Influencing Independence in Adolescents With Sickle Cell Disease	Quantitative	74 AYA	Outcome/Individual/Family support	Factors that predicted independence in adolescents were Knowledge, severity, & family relationships, but accounted for only 25% of the variance
Stollon et al. (2015)	Transitioning Adolescents and Young Adults With Sickle Cell Disease From Pediatric to Adult Health Care: Provider Perspectives	Qualitative	13 providers	Outcome/Individual/Family support/health care system	Transition success was measured by health care utilization, quality of life, and continuation on a stable disease trajectory. Barriers to transition: negative experiences in the emergency department, sociodemographic factors, and adolescent skills. Facilitators: positive relationship with the provider, family support, developmental maturity
Andemariam et al. (2014)	Identification of Risk Factors for an Unsuccessful Transition from Pediatric to Adult Sickle Cell Disease Care	Quantitative	47 AYA	Outcome/Individual	32% did not transition successfully. Gender, race, insurance type, & clinic attendance rate did not influence transition outcome. Travel distance to the adult SCD center, older age at first combined transition clinic visit, & milder disease severity were a significant risk factor unsuccessful transition

Latzman et al. (2010)	Transitioning to adult care among adolescents with sickle cell disease: a transitioning clinic based on patient and caregiver concerns and needs	Quantitative	71 AYA & their caregivers	Outcome/ Health care system	Concerns about transition for pre-transitioning patient included meeting adult providers, while their caregivers were concerned about leaving behind their previous doctor. For transitioning patients and those who already transitioned, the number one concern was being seen in the adult emergency room. Compared to pre-transitioning and patients who have transitioned, patients who are transitioning had significantly lower levels of negative affect and higher levels of positive affect.
Hankins et al. (2012)	A Transition Pilot Program for Adolescents With Sickle Cell Disease	Quantitative	34 adolescents & their parents	Outcome/ Health care system	Program was feasible, and most participants were able to establish an adult medical home
Sobota et al. (2016)	Development of quality indicators for transition from pediatric to adult care in sickle cell disease: A modified Delphi survey of adult providers	Quantitative	79 providers	Outcome/ Individual/ Health care system	Providers chose communication between pediatric and adult providers, timing of first adult visit, patient self-efficacy, quality of life, and trust with their adult provider as top 5 quality indicators for transition in SCD.
Blinder et al. (2015)	Age-related emergency department reliance in patients with sickle cell disease	Quantitative	3,208	Outcome	Transitioning AYAs relied more on the emergency department for care compared to children
Hamideh et al. (2013)	Sickle Cell Disease Related Mortality in the United States (1999–2009)	Quantitative	5416	Outcome	Compared to 1979–1998, the 1999–2009 mortality during childhood has decreased significantly while mortality during the transition age has increased
Blinder et al. (2013)	Age-Related Treatment Patterns in Sickle Cell Disease Patients and the Associated Sickle Cell Complications and Healthcare Costs	Quantitative	3208	Outcome	Compared to pediatric patients, transitioning AYAs had less transfusions, less hydroxyurea & iron chelation therapy, more frequent SCD related complications, and had higher healthcare costs.

Panepinto et al. (2012)	Concentration of Hospital Care for Acute Sickle Cell Disease-Related Visits	Quantitative	13,533	Outcome	Adults were more likely to use multiple hospitals for acute care needs
Dickerson et al. (2012)	Young Adults With SCD in US Children's Hospitals: Are They Different From Adolescents?	Quantitative	18299 adolescents 7072 young adults	Outcome	Compared to adolescents, young adults had higher median admissions per patient per year and higher costs per admission, but similar reasons for admissions and length of stay. Mortality rate was similar between the two groups.
Mennito et al. (2014)	Adolescents with Sickle Cell Disease in a Rural Community: Are They Ready to Transition to Adulthood?	Quantitative	18 AYA	Individual/Health Care Systems	Barrier to transition identified included: leaving their pediatrician, disease knowledge, finding an adult physician, arranging for transportation, preparing to use the adult hospital, keeping a job, knowing what to expect, and handling financial issues. The majority did not have discussions about transition with their provider.
Matthie et al. (2016)	Perceptions of young adults with sickle cell disease concerning their disease experience	Qualitative	29 AYA	Individual/Health care system	Participants reported struggles to maintain or achieve good quality of life, interruptions to family, work and social roles, and difficulties accessing needed health care. They engaged in several strategies to maintain self-care
Griffin et al. (2013)	Applying a Developmental-Ecological Framework to Sickle Cell Disease Transition	Quantitative	135 AYA	Individual/Health care system	Compared to older adolescents, younger adolescents had poorer knowledge of SCD and ineffective management strategies. Program increased knowledge scores over time.
Mulchan et al. (2016)	Applicability of the SMART Model of Transition Readiness for Sickle-Cell Disease	Mixed	14 AYA, 10 clinical experts	Individual/Family support/Health care system	Factors influencing the transition process: relationship with providers; disease-related knowledge; self-management skills; developmental maturity & psychosocial functioning; and beliefs/expectations about transition. Clinical experts tended to identify factors less amendable to intervention such as sociodemographic, while AYAs tended to report more amendable factors such as provider communication

Kayle et al. (2016)	Challenges in Shifting Management Responsibility From Parents to Adolescents With Sickle Cell Disease	Qualitative	14 adolescents 15 parents	Individual/ Family support	Adolescents' adaptive challenges included mastering complex symptom management, communicating about SCD and symptoms, and maintaining control. Parents' adaptive challenges included giving over the complex management, communicating the management with the adolescent, balancing protection against risk with fostering independence, changing a comfortable rhythm, and releasing the adolescent into an "SCD-naive" world. Adolescents' adaptive work included pushing back at parents, defaulting back to parental care, stepping up with time, learning how SCD affects them, and educating friends about SCD. Parents' adaptive work included engaging the adolescent in open dialogue and co-managing with the adolescent.
Porter2014	Transition From Pediatric to Adult Care in Sickle Cell Disease: Perspectives on the Family Role	Qualitative	34 (11 adolescent, 11 sibling, 12 caregiver)	Individual/ Family support	Adolescents, siblings, and caregivers were aware of transition and adolescent's need for more disease management responsibility. Siblings' and caregivers' were concerned that the adolescent might not be able to take care of themselves. Caregivers were concerned about leaving the pediatric environment and adult providers' lack of knowledge. Families recommended more transition preparation opportunities.
Smith et al. (2011)	Growing Up With Sickle Cell Disease: A Pilot Study of a Transition Program for Adolescents With Sickle Cell Disease	Quantitative	33 adolescents	Health care system	knowledge needs for adolescents transitioning to adult care included: ethnicities affected by SCD and inheritance of SCD. Concerns included transferring to a new medical team and they had both positive and negative emotions that varied over time
Tuchman et al. (2008)	Transition to adult care: experiences and expectations of adolescents with a chronic illness	Qualitative	22 AYA (7 with SCD)	Individual / Health care system	Concerns about transition included feeling unprepared and concerns about re-establishing relationships with a new team. Suggestion to improve the process included earlier discussions about transition, meeting the adult team and visiting the adult clinic before transfer.
While et al. (2004)	Living with sickle cell disease the perspective of young people	Qualitative	11 AYA	Health care system	SCD symptoms were complex and taxing requiring various health care service expertise

Telfair et al. (2004)	Transition to adult care for adolescents with sickle cell disease: results of a national survey	Quantitative	172 AYA	Individual / Health care system	Concerns of AYAs were: lack of information related to transition, fear of leaving a familiar provider, fear that the adult provider might not understand their needs, belief that SCD programs should focus on provider support, information about adult care programs, ways to meet adult providers and ways to help adult providers understand their needs
Hauser et al. (1999)	Transitioning adolescents with sickle cell disease to adult-centered care	Qualitative	22 adolescents & 22 parents, 8 providers	Health care provider	Congruence between adolescent and parent concerns about transition; leaving a familiar setting and provider, transferring to an adult provider who may or may not be familiar with SCD, and establishing new family roles. Provider reported barrier to transition included: family and AYA are overly dependent on the pediatric provider, pediatric providers foster dependency, lack of communicating between pediatric and adult providers, and lack of insurance coverage
Tetfair et al. (1994)	Transfer as a component of the transition of adolescents with sickle cell disease to adult care: adolescent, adult, and parent perspectives.	Quantitative	96 AYA, 25 caregivers	Individual / Family support/ Health care provider	Concerns of AYAs payment for medical care and how they would be treated by adult providers. Caregivers were concerned about leaving pediatric care and about adolescent assuming responsibility for their own care. Age, educational level, and disease severity influenced feelings and concerns about transition.
Labore et al. (2017)	Exploring Transition to Self-Management Within the Culture of Sickle Cell Disease	Qualitative	12 AYA	Individual / Family support	Maternal involvement in care was viewed positively by participants; participants described their experiences on growing up in the hospital and learning to manage their condition
Speller-Brown et al. (2015)	Measuring Transition Readiness: A Correlational Study of Perceptions of Parent and Adolescents and Young Adults with Sickle Cell Disease ¹	Quantitative	60 AYA & 60 caregivers	Individual / Family support	AYA's age, amount of responsibility AYAs assume for their healthcare & the degree of parent involvement were factors associated with perceptions of increased readiness to transition

Treadwell et al. (2016)	Development of a sickle cell disease readiness for transition assessment	Quantitative	113 AYA	Individual	Development a SCD specific readiness for transition assessment tool; major areas include: Independent living, knowledge and skills, education and vocation planning, social support skill set
Zhao et al. (2016)	Assessment of Personal Medical History Knowledge in Adolescents with Sickle Cell Disease: A Pilot Study	Quantitative	68 AYA	Individual	Participants were knowledgeable about most of their medical history; emphasizes the importance of tailoring education based on needs
Williams et al. (2016)	Patient-centered Approach to Designing Sickle Cell Transition Education	Quantitative	37 AYA	Individual	Few AYAs receive proper education on transition and the majority expressed interest in learning more about transition
Treadwell et al. (2016)	Self-efficacy and readiness for transition from pediatric to adult care in sickle cell disease	Quantitative	113 AYA	Individual	Self-efficacy was positively associated with readiness to transition. Older age was independently associated with higher scores on the Independent Living Skills scale and higher stress levels were independently associated with lower scores on Education/Vocation Planning scale.
Abel et al. (2015)	Transition Needs of Adolescents With Sickle Cell Disease	Quantitative	122 AYA	Individual	Age and presence of cerebrovascular injury had a significant effect on percentage of participants who reported that they need more practice on areas for transition. The areas with highest percentage of need practice are: living arrangements, money management, vocational skills, and health care skills.
Sobota et al. (2014)	Self-reported Transition Readiness Among Young Adults With Sickle Cell Disease	Quantitative	33 AYA	Individual	Majority reported good SCD knowledge and were motivated to pursue a career despite the burden of SCD. knowledge gaps were in the following areas: independent living & health benefits skills sets.
McPherson et al. (2009)	Transition of Patients With Sickle Cell Disease From Pediatric to Adult Care: Assessing Patient Readiness	Quantitative	70 AYA	Individual	Mean readiness scores were low, Factor associated with transition readiness included age, disease severity, and gender.

Anie et al. (2005)	Multi-site study of transition in adolescents with sickle cell disease in the United Kingdom and the United States	Quantitative	224 AYA	Individual	Demographic variables including age, educational level were associated with physical and psychological symptoms & self-management. Self-efficacy was not associated with demographic factors, but was predicted by physical symptoms,
Sobota et al. (2011)	Transition from pediatric to adult care for sickle cell disease: Results of a survey of pediatric providers	Quantitative	30 centers	Health care system	Majority of centers surveyed had a young transition program. Specific transition practices varied by center.
Rouse 92011)	Informing choice or teaching submission to medical authority: a case study of adolescent transitioning for sickle cell patients	Qualitative	1 counselor	Health care system	Teaching adolescents about the medical culture improved their relationship with the health care system
Okumura et al. (2008)	Comfort of General Internists and General Pediatricians in Providing Care for Young Adults with Chronic Illnesses of Childhood	Quantitative	2434 providers	Health care system	Less than half of general internists felt that their specialty should take primary care responsibility for adult patients with SCD.
Telfair et al. (2004)	Providers' Perspectives and Beliefs Regarding Transition to Adult Care for Adolescents with Sickle Cell Disease	Quantitative	227 providers	Health care system	Adult and pediatric providers agreed that transition programs are necessary. Demonstration of transition included ceasing to see patient, encouraging patients to accept more responsibility, making the patient more financially responsible, and having family conference to discuss transition. Providers who saw only adult patient had different expectations from those who saw both adolescent and adult patients

The Individual domain was one of the most studied domains. Overall, there is evidence that indicates that with increasing age, AYAs report improved transition readiness, yet delaying transition beyond age 21 might be problematic to transition success. However, more research is needed to investigate the appropriate age and/or the appropriate timing for the transfer of AYAs with SCD to adult care. Also, more research is needed to better understand the effect of other demographic factors including gender, socioeconomic status, insurance type, race and ethnicity, educational level, and employment on health care transition in AYAs with SCD. We also found there seems to be a consensus among providers that cognitive deficits might influence transition readiness in AYAs with SCD. However, more research is needed to longitudinally examine the association of disease severity and disease complications, including disabilities, on transition readiness and transition outcomes in AYAs with SCD.

Findings from the family/ social support domain indicated that parents were heavily involved in the AYA's disease management. Although this involvement was viewed as supportive by the AYAs as well HCPs, it's not clear how parental involvement and the parent-AYA relationship can be utilized to facilitate health care transition for the AYAs with SCD and future research should focus on exploring the utilization of the parent-AYA relationship to foster the AYA's readiness and success in transition. More research is also needed to understand the role that social support outside of the family structure plays in the AYA health care transition.

Finally, there were several studies that examined the health care system domain. Findings indicated that, although found to be suboptimal, the AYA-provider relationship in adult care was considered essential for successful transition. AYAs made several recommendations to improve the relationship with the adult provider including introduction to the team before transfer occurs. Findings also indicated that AYAs felt they were not appropriately prepared to transition and had several concerns about the process. They also expressed needs that should be addressed by transition programs to make their transition process easier. Areas for future research include exploring the needs of AYAs who are transitioning to adult care further, identifying interventions that would improve the AYA-provider relationship, and identifying outcome indicators to evaluate the effectiveness of transition programs.

We did not identify any study that focused on the environment domain, indicating that this is an area of need for research. However, we did identify two additional areas that were not explicitly stated in the model: transition success and health care utilization. Although we believe that transition success is an implicit assumption in the outcomes of the model, research exploring how transition success can be operationally defined can be useful to guide the transition of AYAs with SCD. The one study that defined transition success described success from the perspective of the HCPs and found that these providers evaluated successful transition based on health care utilization, quality of life, and continuity on a stable disease trajectory (Stollon et al., 2015). More research is needed to explore the meaning of transition success from

the perspective of the AYAs with SCD and their families. Research is also needed to identify specific and measurable indicators that would allow for the evaluation of transition success in the AYA with SCD.

Although not a component in the HCT model, health care utilization is an important indicator of health care transition and was examined in few of the studies included in this review. The few studies that examined health care utilization during transition indicated that AYAs might at risk of higher acute care utilization, tending to utilize emergency department and hospital care instead of ambulatory care for their health care needs. Co-utilization across services and the factors contributing to the utilization patterns of AYAs with SCD during transition are poorly understood and warrant further research.

2.5 Conclusions

The purpose of this integrative review was to provide a comprehensive review of current state of the science on health care transition for AYAs with SCD. We used the health care transition (HCT) model (Betz et al., 2014) and found the model to be useful in organizing findings. Most studies included in this review addressed the individual and/or health care system domains, few studies addressed the family/ social support domain and HCTs outcomes, and no studies addressed the environmental domain. We also identified transition success and health care utilization as additional domains that were not clearly addressed in the HCT model and discussed areas for future research.

3. Challenges in Shifting Management Responsibility from Parents to Adolescents with Sickle Cell Disease

Annually in the United States, around 750,000 children with chronic illnesses and/or disabilities transition from pediatric to adult health care (AHRQ, 2005). Essential to their readiness for transition is their ability to independently self-manage their condition (Kieckhefer & Trahms, 2000; White, McManus, McAllister, & Cooley, 2012). According to Vygotsky (1978), child cognitive development is fostered within a socially mediated dialectical process in which more competent adults engage children in intentional and meaningful interactions to achieve a higher level of functioning. In the context of children with chronic illnesses, the family constitutes an influential social medium where parents serve as the experienced and competent adults providing the scaffolding (Vygotsky, 1978) that adolescents need in order to become more skilled at self-management. While parents assume the primary responsibility for managing their child's condition during childhood, with time parents need to progressively shift a greater proportion of that responsibility to the adolescent in preparation for their transition to adult health care (Sawyer & Aroni, 2005; Schilling, Knafl, & Grey, 2006; White et al., 2012). Adolescence is a critical period in development where autonomy and personal identity are emerging and, therefore, the progressive shifting of chronic illness management responsibility to the adolescent not only improves self-management skills, but also enhances a sense of responsibility and personal control (Blum et al., 1993; Vygotsky, 1978).

Few studies have examined the shifting of management responsibility from the parent to the adolescent with chronic illness and most of these studies were conducted on parents and

adolescents with diabetes (Allen, Channon, Lowes, Atwell, & Lane, 2011; Karlsson, Arman, & Wikblad, 2008; Leonard, Garwick, & Adwan, 2005; Schilling et al., 2006; Sullivan-Bolyai et al., 2014; Weinger, O'Donnell, & Ritholz, 2001). Findings from this limited body of literature indicate that the shifting of management responsibility is a complex adaptive process for the adolescent and the parent, both of whom need to adjust to the changes in their roles (Allen et al., 2011; Chilton & Pires-Yfantouda, 2015; Karlsson et al., 2008; Leonard et al., 2005; Sullivan-Bolyai et al., 2014).

Adolescents need to progressively switch roles from being dependent on parental management to functioning as leaders in their own care. Building the capacity to take the lead in the self-management of their conditions requires that adolescents develop additional skills in problem solving, decision making, knowledge about their condition, along with mastery of the relevant technical self-care skills (Bell & Sawyer, 2010; Karlsson et al., 2008; Sawyer & Aroni, 2005).

Concurrently, parents need to gradually relinquish their role as the primary managers of care to the role of advisors and continuing supporters of the adolescent (Allen et al., 2011; Karlsson et al., 2008; Sullivan-Bolyai et al., 2014). While some parents described being able to back off either partially or completely to allow more independent adolescent self-management (Sullivan-Bolyai et al., 2014), other parents described being anxious and uncertain about the extent of the responsibility they can relinquish to the adolescent (Buford, 2004; Sullivan-Bolyai et al., 2014). The parental struggle to progressively relinquish control to the adolescents is often

due to fear of worsening health outcomes (Buford, 2004; Hanna & Guthrie, 2000; Sawyer & Aroni, 2005), especially if parents believe that the adolescents are not as competent as they are in managing their condition (Allen et al., 2011). Shifting towards more independent adolescent self-management might also create conflict between the adolescent and parents as they often disagree on management decisions and long-term health goals (Buford, 2004; Weinger et al., 2001).

While these studies emphasized the challenging nature of shifting the management responsibility from the parents to the adolescent, little has been done to describe specific challenges in depth using the perspectives of adolescents and their parents. A better understanding of these challenges is warranted, particularly in other health care conditions.

In this study we explored the challenges of progressively shifting management responsibility from parents to adolescents with Sickle Cell Disease (SCD). SCD is an ideal condition to study the shifting of management responsibility because of its complexity and unpredictability and the resulting self-management challenges it imposes on individuals and their families. Individuals with SCD suffer from a variety of complications including pain crises, chronic pain, acute chest syndrome, and stroke (Miller & Meier, 2012). The symptoms and complications that accompany this complex chronic condition occur across an unpredictable trajectory that varies among individuals and within the same individual over time (Miller & Meier, 2012). Self-management of SCD entails monitoring symptoms on a daily basis; adhering to aggressive lifelong disease management, including hydroxyurea, chronic transfusion, iron

chelation, and pain management; as well as preventive management such as hydration and rest (Miller & Meier, 2012).

Adolescence is a particularly vulnerable period for individuals with SCD. Compared to younger children with SCD, adolescents have worse health outcomes including higher rates of complications (Darbari et al., 2012; Lanzkron, Carroll, & Haywood, 2013; Quinn, Rogers, McCavit, & Buchanan, 2010); higher rates of depression and anxiety (Benton, Ifeagwu, & Smith-Whitley, 2007; Jerrell, Tripathi, & McIntyre, 2011); higher utilization of care (Brousseau, Owens, Mosso, Panepinto, & Steiner, 2010; Fosdal & Wojner-Alexandrov, 2007; Lotstein, Inkelas, Hays, Halfon, & Brook, 2008); and higher risk for mortality (Hassell, 2010; Lanzkron et al., 2013; Quinn et al., 2010). Improving health outcomes for this age group mandates assisting adolescents to increasingly engage in self-management (Aujoulat et al., 2014; Sattoe, Hilberink, Peeters, & van Staa, 2014).

Further, SCD is a genetically inherited condition with manifestations occurring in infancy, resulting in the parents' assuming the role of the primary manager of care from the child's birth. By the time their child reaches adolescence, parents have been in the role of the primary manager of care for more than a decade. Shifting management to the adolescent might pose a unique challenge for parents for whom the care management role has become part of their parental identity.

Importantly, individuals with SCD face racial stigma (Institute of Medicine, 2003) as well as disease stigma (Haywood, Tanabe, Naik, Beach, & Lanzkron, 2013). For example,

adolescents with SCD reported unmet health care needs where they felt their pain is ignored and misunderstood as drug-seeking behavior (Erskine, 2012; Telfair, Alexander, Loosier, Alleman-Velez, & Simmons, 2004). Parents reported that they often find themselves obliged to negotiate and advocate on behalf of their children during health care encounters and in their communities (Dyson, Atkin, Culley, Dyson, & Evans, 2011; Graff et al., 2010; Mitchell et al., 2007). These experiences might intensify the role parents play in their adolescents' health, leaving little room for adolescent self-management (Brown, Connelly, Rittle, & Clouse, 2006; Oliver-Carpenter, Barach, Crosby, Valenzuela, & Mitchell, 2011; While & Mullen, 2004).

Very few studies examined self-management in adolescents with SCD (Andemariam et al., 2014; Jordan, Swerdlow, & Coates, 2013; Labore, Mawn, Dixon, & Andemariam, 2015; Newland, 2008; Telfair, Ehiri, Loosier, & Baskin, 2004; Telfair, Myers, & Drezner, 1994; Treadwell, Telfair, Gibson, Johnson, & Osunkwo, 2011) and none directly addressed the challenges of shifting of management responsibility from the parent to the adolescent with SCD (Sawyer, Drew, Yeo, & Britto, 2007). As a result, there is a limited understanding of the shifting of the management responsibility from the parent to the adolescent with SCD.

3.1 Theoretical framework

The Adaptive Leadership Framework for Chronic Illness (Anderson et al., 2015) provides useful direction for the study of challenges in shifting management responsibility from the parents to the adolescent with SCD. The framework distinguishes between technical and adaptive types of challenges and draws attention to the collaborative work between the patient,

family, and provider necessary to arrive at a shared meaning of the challenges and solutions for increasing the adaptive capacity of both adolescent and parent (Thygeson, Morrissey, & Ulstad, 2010). Technical challenges are clearly defined problems best addressed with clinical expertise, usually of the provider using medical interventions such as prescribing medications. Adaptive challenges are not easily defined, involve many interrelated factors, and require the adolescent/family to do the work required to adapt to a new reality (Thygeson et al., 2010). For example, the use of pain medications constitutes technical work used to address the challenge of pain, while integrating medication management in the adolescent's daily routine at school constitutes an adaptive challenge. Initially, the adolescent might lack the skills or resources to do the adaptive work and thus learning new behaviors is needed to enhance adaptive capacity. The provider's role is to help adolescents and their parents to identify challenges and develop the needed skills to do adaptive work.

The adaptive/technical distinction is very useful when parsing the self-management needs of adolescents with SCD as these adaptive challenges are often on their minds, though rarely addressed in typical clinical encounters (Mitchell et al., 2007). The complexity and unpredictability of SCD (Miller & Meier, 2012) imposes the need for technical as well as adaptive work and the use of collaborative approaches by the adolescent, parent, and provider (Labore et al., 2015). Given the importance of developing independence in self-management for improved health outcomes, research is needed to understand the challenges that adolescents and parents have in progressively shifting the management responsibility from the parents to

the adolescent (Sawyer & Aroni, 2005). Therefore, the purpose of this study was to explore and describe the challenges faced by adolescents with SCD and their parents and the work they engage in to progressively shift from parent management to independent adolescent self-management.

3.2 Design and Methods

3.2.1 Study design

A qualitative, descriptive focus group design was used for this study. Focus groups are useful for gaining in-depth understanding of the participants' perceptions of and experiences with health-related issues. In particular, focus groups are useful and feasible for studying adolescents' perceptions and experiences regarding health-related issues because they acknowledge the adolescents as the experts on their condition (Gibson, 2007; Heary & Hennessy, 2002). Further, the group interaction in focus groups allows participants to react to and build on each other's responses, resulting in richer data that might have not been produced otherwise with individual interviews (Stewart, Shamdasani, & Rook, 2007).

3.2.2 Participants

Study participants were recruited by provider- and self-referral from a major pediatric sickle cell center in the southeast United States. Adolescent inclusion criteria were: a) SCD (HbSS genotype), b) 11-18 years, c) English speaking, and d) ability to provide consent/assent. Parent inclusion criteria were: a) primary caregiver of an adolescent with SCD, b) daily interaction with the adolescent, c) English speaking, and d) ability to provide consent.

Grandparents and other guardians were eligible to participate as the parent participant if they were the primary caregiver and the guardians for the adolescent. Demographic information for all participants and self-report health information for the adolescents were collected. A semi-structured interview guide with probes was used for the focus group interviews.

Two adolescent focus groups, with a total of 14 adolescents, and two parent focus groups with a total of 15 parents were recruited, convened, and interviewed. Adolescent demographic characteristics and disease characteristics and parent demographic characteristics are summarized in Tables 1 and 2, respectively. The average adolescent participant was 14.4 years, male, African American, had an average of 3.2 pain crises in the last two years, and was on hydroxyurea and pain medications. The average parent participant was 44 years, female, African American, employed, and had a college degree. Parent participants included eleven biological parents, one adoptive parent, two grandmothers, and one godfather.

3.2.3 Data collection instruments and procedures

After Institutional Review Board approval was granted, written informed consent was obtained from all parents or guardians and 18-year-old adolescents. Written assent in addition to parental consent was obtained for adolescents younger than 18 years. Adolescent and parent focus groups were conducted concurrently in two separate rooms on two occasions. While focus group methodology has been found to be a useful method for research involving adolescents and parents (Gibson, 2007; Heary & Hennessy, 2002; Stewart, Shamdasani, & Rook, 2007) there remains the possibility that some group members may find talking in a group

format intimidating while others might be dominating the discussion. To offset this potential problem, we ensured that the focus group facilitator attempted to control and enable opportunities for participation across members of the focus group. Facilitators were MSN prepared nurses trained in focus group methodology. One facilitator had adult nursing experience and conducted the parent focus groups and one facilitator had extensive pediatric nursing experience and conducted the adolescent focus groups. In our study, the group process seemed to work well as participants engaged actively within the group and frequently built on each other's responses. Some of our adolescent participants were siblings and some of the adolescents and parents were acquainted with each other outside the focus group, which might have also facilitated participation for the adolescent and parent focus groups.

Focus group discussion guides included the following topical areas: adolescent challenges with managing SCD and adolescent's work for managing SCD for the adolescent focus groups, and parents' challenges with managing SCD and parents' work for managing SCD for the parent focus groups. For example, adolescents were asked about their challenges with managing SCD by posing the grand tour question, "Tell me about the difficulties you have with taking care of your sickle cell". When needed, a probing question such as "tell me more about that" was used. Following the interviews, participants completed the demographic form. Data collection ranged between 60-90 minutes. Participants received a \$50 gift card, lunch during the focus group, and transportation reimbursement.

After each focus group, the two facilitators met, shared, and discussed field notes. The digital recordings were transcribed verbatim into an electronic transcript. Transcription was conducted by an experienced and well-trained transcriptionist, and transcripts were proofed against the audio recordings by the first author (MK) who conducted the adolescent focus groups. To maintain confidentiality, pseudonyms were used for study participants.

3.2.4 Data analysis

Transcripts were analyzed using content analysis (Bernard & Ryan, 2010; Hsieh & Shannon, 2005). Parent focus groups transcripts and adolescent focus groups transcripts were coded and analyzed separately. Digital transcripts were viewed using ATLAS.ti 6 (Scientific Software Development GmbH, Berlin, 2011), a text analysis program. Each transcript was read to get a general feel for the data and incorporate field notes were incorporated. Next, transcripts were coded. Provisional codes based on the Adaptive Leadership Framework for Chronic Illness (Anderson et al., 2015), namely technical and adaptive challenges and technical and adaptive work, were used as classification categories. Open coding was then used to identify challenges and work. This integrated approach of using deductive and inductive coding (Bradley, Curry, & Devers, 2007) allowed the researchers to go beyond the general concepts in the framework. A code book was developed and iteratively refined throughout the analysis and included codes, field notes, and evolving ideas. The first author (MK) developed the initial code set and applied them to 50% of the transcripts. A second coder (SD), with extensive coding experience, reviewed the initial 50% of coding and consensus was used to refine the codes and

arrive at 100% agreement on applied codes. Once all transcripts were coded, categories were identified for parent focus groups and adolescent focus groups across codes by searching for patterns and links, variation in data, consequence, and context (Richards, 2013) within each focus group type (parent and adolescent focus groups). The salience of particular codes was determined based on frequency; pervasiveness, similarities and differences across focus groups and the relation to the framework (Bernard & Ryan, 2010). Codes were abstracted into a matrix that included focus group characteristics, abstracted text, code, category, and relevant context. Trustworthiness and rigor of the analysis procedure was constantly assessed and supported by the use of a systematic coding procedure employing two coders, an audit trail of all analysis decisions, and detailed description of each code and category.

3.3 Results

When asked about the challenges they have with SCD and the work they do to manage it, both adolescents and parents predominantly discussed challenges and work that were adaptive in nature. Neither group described challenges or work that would be categorized as technical.

3.3.1 Adolescent adaptive challenges

Adolescents described three classes of adaptive challenges to self-manage SCD: Mastering complex symptom management, communicating about SCD and symptoms, and maintaining control. These challenges were related to their condition and how it affected their lives as adolescents.

Mastering complex symptom management. The adolescents discussed the challenge of managing the complex symptoms associated with SCD. Although they discussed several self-care strategies, they still struggled with managing the unpredictability of the symptoms.

14-year-old female: "It's difficult because you don't know when or what's going to happen, sometimes it's going to hurt your chest; sometimes you get a really bad headache. You just don't know what to do."

Further, the adolescents discussed how because of the unpredictable symptoms, SCD interrupted their lives and they missed out on significant events. The unpredictability of symptoms impacted socialization with peers and was seen as one of the most distressing aspects of symptoms.

16-year-old male: "I was nominated for Mr. Freshman, and then I had a pain crisis. So I couldn't go to the dance. It just messed up everything and it just came at the most random times. You don't feel an initial pain or an initial sense, it just happens to you."

Communicating about SCD and symptoms. The adolescents felt challenged in communicating to others; including parents, peers, teachers, and providers about their SCD and associated symptoms. Some preferred to manage symptoms alone rather than seek help from a parent. They also preferred to manage symptoms at home, rather than seek help from a provider. This challenge resided in the nature of SCD symptoms, such as pain crises, which are subjective and not visible to others.

The adolescents expressed a particular frustration with having to explain over and over again to teachers and peers that they had SCD, how SCD affected them and what they needed to do to prevent and manage symptoms.

14-year-old male: "Sometimes they [coaches and teachers] forget and you'll be running the mile and they'll see you slowing down and they'll pressure you 'go, go' and you have to remind them that you still have sickle cell, it doesn't go away. They thought it would go away."

Further, the adolescents discussed how they were selective with whom they disclosed having SCD and chose to tell a small group of trusted friends and teachers.

15-year-old female: "I will tell some [friends], but I don't tell males I have sickle cell. Partly because I told one before and that changed our whole relationship."

14-year-old female: "I don't talk to nobody about it because I don't want them to feel sorry for me because I have sickle cell. I just don't talk to people about it except my mom, my parents, my brothers."

Maintaining control. Because of the unpredictable nature of the symptoms and their incapacitating effect, the adolescents struggled with maintaining control over what happened to them. SCD disrupted normality and their daily life, leaving them little control over what activities they can engage in. This lack of control left them feeling stuck with SCD for a lifetime with few options available.

15-year-old female: "It's hard because I can have pain at any time and last for maybe a week, two weeks. I hate taking my hydroxyurea because I have to take it for the rest of my life. I

could get the bone marrow transplant I could die and my mom really doesn't want me to. So I'm just going to have to deal with it. And I just want to be like every other kid that doesn't have sickle cell. It's hard dealing with it but it won't go away. I wish it would but it won't."

3.3.2 Adolescent adaptive work

The adolescents engaged in several types of adaptive work to address the challenges associated with more independent self-management including: Pushing back at parents, defaulting back to parental care, stepping up with time, learning how SCD affects them, and educating friends about SCD. These strategies were aimed towards achieving more autonomy for the adolescent.

Pushing back at parents. One strategy that adolescent used to address the challenges of acquiring more independent self-management was to push back at parents for more control. Adolescents described situations where parents were perceived as overprotective with care, described their frustration with the overprotection, and expressed the need for more autonomy.

16-year-old male: "And now that I'm older, when I go out with my friends, she's like, you need to drink water. Mom I know! I'm sixteen years old I'm going to be an adult soon. Gosh! So maybe if my mom would give me a little bit more space, then maybe I know all this stuff."

The adolescents were also frustrated with parents taking over the management and pushed back for more decision making control.

15-year-old male: “[Don’t ask] what number are you at [pain score]! [Ask] do you need medicine, is the medicine working? Your five might be different than my five so how do you judge for me. By this point I pretty much know my body and how it works.”

Defaulting back to parental care. Despite pushing back at parents, the adolescents described how they defaulted back to parental care when they could not control their symptoms. According to the adolescents, parents were trusted caregivers who knew exactly what to do in these situations. The adolescents seemed to default back to the parent for comfort and support as well as complex symptom management.

14-year-old female: “My mother helps me a lot and she will know when I have a crisis, it’s just instinct. So, I go to my mom every time I have a crisis because she knows how to handle it really well than anybody else. Every parent in here knows what’s up because they know what to do.”

Stepping up with time. The older adolescents explained to the younger adolescents in the group how they stepped up to take more responsibility of their care as they got older. Their taking on more responsibility included taking medications and scheduling clinic appointments on their own, with little interjection from parents. Parents still checked on their progress, but the adolescents took a more proactive role in their management.

18-year-old female: “Once you get older it’s like ‘oh wait I’m different, I really have to take care of myself.’ You feel more responsible.”

17-year-old male: "As you get older you have to try to take more responsibility for taking care of yourself. You said this is my mom's job. That's not bad but at the same time you want to know this stuff because if you have to go to the hospital, you need to be able to answer questions because if they have to wait for your mom they can't do anything until she gets there."

Learning how SCD affects them. This strategy was also particularly evident with the older adolescents in the group. The adolescents discussed how they were attuned to their symptoms and were trying to understand how SCD affected them so that they could attempt to prevent pain crises. The adolescents discussed the self-care strategies they used to prevent pain crises, such as resting when tired and taking pain medications and hydrating before engaging in activity.

17-year-old male: "What I always do before running I take Ibuprofen and keep a water bottle with me. I know I'm going to be doing a lot of physical stuff, I'm going to go ahead and take this pain medicine."

Educating friends about SCD. The adolescents discussed how they strategically educated friends about their condition. Again, this strategy was evident among the older adolescents and appeared to serve two purposes: fitting in with peers and ensuring peer support. Educating friends about SCD and how it affected them appeared to help the adolescents with their relationship with friends, because friends, for example, understood their absences during hospitalization. Further, friends who knew how SCD affected them could be a source of support for the adolescent during pain crises.

16-year-old male: "I'd rather them [friends] know about what I have so that if I'm not here for a week they're not thinking I'm just ditching them. They kind of know my routine."

17-year-old male: "Your friends will try to take care of you. Make sure you're with people that if something happens then they'll help you, do what needs to get done if you can't yourself."

3.3.3 Parent adaptive challenges

Parents' adaptive challenges included giving over the complex management, communicating the management with the adolescent, balancing protection against risk with fostering independence, changing a comfortable rhythm, and releasing the adolescent into a "SCD-naive" world. The parents' challenges were related to the complexity of SCD, lack of public understanding of SCD, the adolescent's development, and their own comfort with the status quo.

Giving over the complex management. Mirroring the adolescents' challenge of taking on responsibility of managing complex symptoms was the parents' challenge of shifting the complex management to the adolescents. Parents felt that they had mastered the complex symptom management for SCD because they were engaged in the management since the birth of their child. Parents described the many ways they had learned to navigate through clinical and knowledge sources, including self-education, professional recommendations, and firsthand experience with family members with SCD. Their management decisions were based on the triangulation of different sources and what they called "parental instinct." They looked at all management advice from providers with a skeptical eye and triangulated different sources of

information to eventually decide on the best course of action for their child. Mentoring the adolescent to master this strategy constituted a challenge for them particularly amidst a disease trajectory that was unpredictable.

Mother of 13-year-old female: "Once you learn that they have [SCD], I looked at every possible outlet. I can give seminars on it. So when I explain to someone what my daughter is dealing with, there is no question you can ask me that I can't answer... it's all for me parent instinct. Because you're giving me this option as an alternative I'm not, my daughter is not a guinea pig."

The parents felt the need to stay in control of the management because they feared that the adolescent might not be able to handle the full complexity of care. The parents described periods in which they were able to step back and allow the adolescent some independence in management, but in general they felt the need to pay close attention to the adolescents' health status with close monitoring and assessment of hydration, diet, and medications.

Mother of a 15-year-old male: "the reason why I feel protective is I think my son won't take care of himself like I would."

Mother of an 18-year-old male and 15-year-old female: "That's my fear because I stay on him. You were healthier when you were younger because I could control you more [others agreeing] ... it's making sure that I'm on it with the medicine, I'm on it for the hydration. Those are my two things, if nothing else I do that day that my child has their medication and he is hydrated and happy and we've made it."

The parents struggled with giving over to the adolescent the monitoring portion of SCD symptom management. They expressed that they have had many years of experience understanding how their adolescent responded to SCD and they had developed the expertise in monitoring and managing symptoms. Paradoxically, they felt that their adolescent lacked this experience.

Mother of a 16-year-old male: "We've had 16, 18 years to learn, to watch them and to see 'okay the eyes are turning yellow, this is happening, that's happening' to learn what goes on with him. He thinks he knows himself. You actually know him better than he knows himself right now. I was the one dragging him to the hospital and I remember all those IVs and crazy chest x-rays; he doesn't!"

Communicating the management with the adolescent. Parallel to the adolescents' challenges of communicating symptoms, the parents also described how adolescents are often reluctant to talk to them about their symptoms and, therefore, managing symptoms became more difficult.

Grandmother of 11-year-old and 13-year-old males: "My eldest grandson tends not to want to go to the hospital so he doesn't let us know when he's in pain, so it makes handling his crisis really difficult."

Mother of 16-year-old male: "I know when he's going through crisis when I get the water bill, or if I wake up and my thermostat is at 90 degrees and he has a heating pad and a blanket on him. 'Why didn't you tell me you are in pain?'"

The parents also expressed difficulty in talking to their adolescent about their care. They were fearful about shifting management responsibility to their adolescent because conflicts often arose. For the parents, communicating their thoughts on disease management at this age was a challenge because the adolescents were often not “receptive” to parental opinion.

Mother of 16-year-old male: “I feel he needs to get to know himself and that’s tough during the teenage years because they’re not real receptive to your parental opinions and advice at this age.”

Mother of 16-year-old adopted male: “It’s kind of a hard balance to make sure they understand these things in a way that they’ll receive it. That’s just the harder part shifting some of that responsibility.”

Balancing protection against risk with fostering independence. The parents described the challenge of balancing protecting the adolescent from SCD complications and allowing the adolescent opportunities to become more independent. The parents worried that the adolescent might be willing to take “extreme” risks for better quality of life and this was a situation that the parents were not comfortable with.

Mother of 18-year-old female: “What broke my heart was somebody put it in her head about the bone marrow transfusion. She asked the doctor, and the doctor was like ‘it’s like a one in ten chance [survival]’. And my little girl actually looked at us and said ‘I’d rather take the chance.’ And I thought- Now that tore me up, I cried. You’d rather risk death than go through what you’re going through!”

Finding the balance between close oversight of the care needs imposed by SCD and allowing the adolescent to explore and experience the world was often very difficult for the parents. The parents talked about feeling torn between fostering growth and independence for the adolescent and preventing complications.

Mother of 18-year-old male: "I remember my first snowfall with him. My oldest son went outside and I won't let him go because I was just so 'Oh my Gosh, he will have a crisis", but he was like 'I'm going to miss everything, I hate these sickle cell.' And I was like 'Oh Gosh, okay, I'm going to layer you off'. And with the sports he didn't like it, which was a good thing for me."

Mother of 14-year-old female: "You never want it to be one of those things where you've restricted them from so much and now they can't [do things]. I have two other kids that are very active in sports. Sometimes I stop her 'no you can't participate in anything' and she'll throw that at me 'Why are you hindering me? You think I'm sick mom!' So now she's doing ballet, she's doing hip hop dance. I'm like 'you know you're doing too much.'"

Changing a comfortable rhythm. The parents described how they and their adolescents live through "pockets of time" because of the unpredictability of SCD. The parents described how across the years they learned how to move between the periods of illness and health and manage the needs of the adolescent. They had developed a "rhythm to living life".

Mother of 18-year-old female and 14 years old male: "what is tricky about sickle cell is it's there every day, but you don't live it every day. It's like pockets of times when you're dealing

with it. Our son just got out of the hospital and re-entering into life is always challenging after a hospital stay. He was like 'I'm afraid it is going to happen again.' ...but you have to look at it like making deposits, you are feeling good, let's make our deposits, when it happens again then we'll make the withdrawals.' And I'm telling myself that! It takes us almost a week to kind of get [back to normal]. That first week we're still dealing with the pain, we're backing it down, so it's like that re-entry into life, it can be bumpy."

Shifting care responsibility to the adolescent was a challenge because it forced the parents to change their rhythm and risk the balance they had achieved and to take on yet another task of mentoring the adolescent in SCD management.

Mother of 14- year-old female: "I got her balanceIt's just that how do you feel separating yourself from your child, knowing that they're going through that. That's the hard part! And she's big now 'mom I can take care of myself'. 'What did you call me? Mom! Exactly! Let me do my job please!'"

Releasing the adolescent into a "SCD-naive" world. The parents felt that there was a lack of awareness about SCD, both in their immediate circle and in the community they live in and that created a challenge for them to trust that the adolescent will receive the support they needed when the parents were not present. Parents shared stories of how their awareness about SCD was through their firsthand experience with it.

Mother of 14-year-old female: "There really hasn't been much that people knew about it until you start having kids, or you know family members that have it. And so many people are

oblivious to the fact that this is serious. We run into it at school, 'oh what is that?' 'What do you mean, what is it? I've known it for the last fourteen years! Why you don't know about it!'"

The parents felt it was necessary for them to advocate for the adolescent and make sure that their needs are met, especially at school.

Godfather of 17-year-old male: "I'm not sure that a lot of people at school understood that he needed to go to that water fountain or restroom, please do not hinder him because I might come back looking."

Mother of 15-year-old female: "When she was in the elementary school, the teacher wouldn't let her go to the bathroom. I said just get up and walk out, [if] they say anything tell them to call your mom. And then the principal called and I said 'I told her to walk out. My daughter has sickle cell, when she needs to get water or whatever, she needs to do it. She isn't going to ask anybody because her mom told her to do it.'"

Mother of 14-year-old female: "That must be our slogan I'll deal with them."

3.3.4 Parent adaptive work

In contrast to the adolescents, parents described less adaptive work to address the adaptive challenges of shifting the management responsibility. Coping with the unpredictable day to day care needs of the adolescent with SCD appeared to be taxing for parents, who, on top of the complex management, need to develop strategies to mentor the adolescent towards independent self-management. The parents' adaptive work included: engaging the adolescent in open dialogue and co-managing with the adolescent.

Engaging the adolescent in open dialogue. One strategy the parents described was engaging the adolescent in open dialogue about SCD and its management. Parents hoped that engaging in these discussions will equip the adolescent with a better understanding of their condition and its management.

Grandmother of 15-year-old male: "We always talk. It's communicating, teaching him, 'I can see that you're not feeling it. Tell me how you feel' and, but now it's mostly I have to remind him 'take your medicine.' He'll drink the water. He's eating better, so I can kind of lean back a little bit."

Co-managing with the adolescent. Another strategy that the parents described was collaborating with the adolescent on SCD management. While some parents would do what is needed, other parents stepped back to allow the adolescent to take on responsibility for their care. Stepping back did not mean that the parents completely let go of management; they still overlooked care and monitored the adolescent management.

Mother of 18-year-old male: "He's responsible for his own medication, but I did step back and just let him, the time that I stepped back I said I've watched you for the last two weeks and you've taken your medicine around five times. I just gave him some tips: 'set an alarm, your phone to remind yourself because I'm not always going to be able to remind you.'"

3.4 Discussion

This study explored the challenges that adolescents with SCD and their parents face and the work they do to shift the management responsibility from the parents to the adolescent. To

our knowledge, this is the first study to examine this shifting work and the challenges that it brings for both adolescent with SCD and their parents.

For the adolescents in our study, complex unpredictable symptoms made it difficult to take on the self-management responsibility. The addition of the non-normative stressors of complex SCD management layered on top of normative developmental challenges and transitions faced by adolescents (Kennedy, Gask, & Rogers, 2005). The adolescents also struggled with maintaining control over the events in their lives because of the unpredictability of SCD. This challenge is particularly significant given that achieving autonomy and independence are important developmental milestones for adolescents. The adolescents addressed this challenge with adaptive work directed at having more autonomy over their management, including understanding how SCD affected them, pushing back at parents and over time stepping up for more decision making responsibility.

Accepting responsibility of care also requires that the adolescents be able to advocate for themselves. The adolescents in our study were reluctant to discuss their condition with their peers and teachers and some did not disclose symptoms to parents and providers. The adaptive work they did to address the challenge of communicating symptoms to others was to educate their friends on SCD and to default back to their parents for disease management and support. Educating friends about SCD helped the adolescents to maintain their social relationships with friends and to tap into their friends' assistance with SCD management when needed. Defaulting back to parents for SCD management served to elicit parental expertise when the situation was

beyond what the adolescents could manage on their own. Parents also served as a source of comfort and support to the adolescents.

The adolescents' struggle with managing complex symptoms was mirrored by parents' hesitation to fully shift the management responsibility to the adolescent, because parents felt that the adolescent lacked their level of expertise in SCD management. Mentoring the adolescent to master disease management and triangulate different knowledge sources as the parents had done over the years constituted a challenge for the parents, particularly amidst an unpredictable disease trajectory. The parents also worried that the adolescent might be willing to take "extreme" risks for better quality of life. This situation made it difficult for them to shift the management decision making to the adolescent because they felt the need to remain in control to ensure the adolescent's wellbeing. Perhaps the greatest challenge for the parents was to balance fostering the adolescents' independence with maintaining their wellbeing. In the case of SCD this challenge comes with a grave cost: hospitalization and sickle cell crises. The parents' adaptive work to address these challenges included engaging the adolescent in discussions about their condition and how they are responding to it as well as collaborating with the adolescent on SCD management.

Interestingly, the adolescents and parents in our study discussed only adaptive challenges and adaptive work and neither discussed technical challenges or technical work. Similarly, Palladino and Helgeson (2013) found that adolescents, parents and physicians agreed that youth with diabetes were more proficient at technical medical skills (e.g. insulin

administration) than adaptive life-style behaviors such as diet and exercise. Chilton and Pires-Yfantouda (2015) also found that acquiring self-management responsibility for adolescents with diabetes was more adaptive in nature. For example, they found that integrating self-management in social contexts such as school was difficult for adolescents with diabetes because they perceived that managing diabetes at school elicited negative attention from peers. These findings along with our findings emphasize the importance of adaptive challenges and adaptive behaviors in self-management and indicate the need for more research focused on assisting adolescents with chronic illness and their parents in addressing adaptive challenges with shifting management responsibility to the adolescent.

Our finding that adolescents had a more proactive role in their care as they got older was not surprising and is consistent with the older adolescent's development of autonomy. Similarly, Labore et al. (2015) found that participants in their study reported taking more control of SCD management between the ages of 16 and 24 years. Findings from the diabetes literature also indicate that adolescents engaged in more self-management with increasing age (Kelo, Martikainen, & Eriksson, 2011; Palmer et al., 2004; Schilling et al., 2006). However, Palmer et al. (2004) found that child's age and physical maturation, rather than their level of competence and autonomy, explained the mother's transfer of responsibility for diabetes management. On the other hand, Buford (2004) found that mothers of children with asthma responded to specific cues, such as the child taking initiatives, to involve their children more in asthma management. Relying on chronological age for shifting management responsibility

might be risky. Age is not congruent with competency, and shifting care responsibility should be based on the adolescent's maturity, self-efficacy, and ability to assume responsibility of care (Young, Lord, Patel, Gruhn, & Jaser, 2014). These findings call for more research focused on assessing the adolescent's readiness and their competency in taking on more management responsibility.

We also found that the parents in our study continued to maintain oversight over the adolescent's management, an approach that was reflected in both the adolescent and the parent focus groups. Other studies have similarly found that adolescents with diabetes hovered between independent self-management and relying on parental decision making, while parents continued to provide support and advice in management decision making (Allen et al., 2011; Karlsson et al., 2008; Sullivan-Bolyai et al., 2014). In two diabetes reviews, parental involvement in care was significantly associated with better adolescent treatment adherence (Dashiff, Hardeman, & McLain, 2008; Young et al., 2014). Productive parental communication characterized by support, involvement, and warmth was associated with better metabolic control and treatment adherence, while problematic parental communication characterized by conflict, negative affect, and control was associated with worse metabolic control, self-care, and quality of life in the adolescent (Dashiff et al., 2008). Similarly, Labore et al. (2015) found that young adults with SCD appreciated maternal involvement in care and viewed maternal involvement as essential to them transitioning to self-management. Balancing monitoring the

adolescent with enhancing the adolescent's self-efficacy should be attempted (Young et al., 2014).

Parent-adolescent communication on disease management was found to be a challenge for both the parents and adolescents in our study. To overcome this challenge, parents engaged the adolescents in open dialogue about SCD, its management, and the adolescent's response to it. Adolescents with diabetes viewed open parental communication that encouraged responsibility, discussed the adolescent struggles, and recognized progress as supportive (Dashiff et al., 2008; Young et al., 2014). Encouraging parent-adolescent communication on disease management might be a useful intervention in building the adolescent's capacity in self-management. Research exploring the feasibility and effectiveness of interventions aimed on facilitating parent-adolescent communication is needed.

A review by Kelo et al. (2011) found that mothers of adolescents with diabetes built supportive networks with teachers, nurses, and peers at school and expected the school to provide support and be involved in diabetes management. In contrast, the adolescents and parents in our study described that teachers and peers did not understand the gravity of SCD and were not supportive of SCD management. Parents described having to aggressively advocate on their child's behalf at the school. Our finding is in line with other research on SCD where parents of children with SCD often find themselves obliged to negotiate and advocate on behalf of their children during health care encounters and in the community (Dyson et al., 2011; Graff et al., 2010; Mitchell et al., 2007). When tailored to the adolescent's need, parental

advocacy was associated with better coping strategies, better utilization of health care services, and decreased anxiety and depressive symptoms; however, this often led to parents taking a predominant role in their child's health (Brown et al., 2006; Oliver-Carpenter et al., 2011; While & Mullen, 2004) and to disagreement between the adolescent and parent about disease management (Burlaw, Telfair, Colangelo, & Wright, 2000; Erskine, 2012).

3.5 Study Limitations

Our study recruitment was limited to one medical center, which might limit the external validity of the study. However, the center from which the participants were recruited is a major referral center in the southeast United States, and participants from a geographical radius of up to 100 miles from the center were recruited to the study. Our adolescent and parent samples were mostly African Americans. However, this is expected given that SCD occurs predominately in African Americans in the United States (Hassell, 2010). Our adolescent sample consisted mostly of male participants, with only three female adolescents. Yet, our adolescent sample was similar to reported population estimates in the mean number of hospital admissions and emergency department use (Paulukonis et al., 2014). Finally, our parent sample had at least a high school education and consisted mostly of female caregivers. The educational level of our parent sample might limit the generalizability of our findings to other parent samples. The fact that the majority of the adolescents' primary caregivers were female was not surprising; studies report on the important role that female caregivers play in the care of the chronically ill in the African American population (Becker, Gates, & Newsom, 2004; Jenerette & Valrie, 2010). Despite these

limitations, this study contributes in-depth understanding of the challenges that adolescents with SCD and their parents face in shifting the management responsibility from the perspectives of adolescents and parents.

3.6 Implications for Practice and Research

Vygotsky emphasized the role of social forces, including parental guidance and interaction, teacher instruction, and language in child development (Miller, 1993). Within this framework, enhancing self-management capacity in the adolescent requires an iterative process of parent-adolescent communication. It might be easier for everyone concerned to maintain the status quo with the parents in full charge of the management, particularly if their strategies have been successful in maintaining the adolescent relatively healthy. While this approach might be effective when the adolescent is residing with the parent, self-management becomes a challenge when the adolescent leaves the home, for example, to college. Health care providers need to collaborate with parents and adolescents to incrementally shift the responsibility of disease management (Young et al., 2014). Health care providers need to assess the parent-child relationship and their progress in shifting the management responsibility, facilitate discussions to arrive on a shared understanding of the challenges they are having with shifting the management responsibility, and collaborate on adaptive work to address these challenges.

Our study described the challenges that adolescent with SCD and their parents face with shifting the management responsibility. Further research should explore the challenges adolescents with other chronic conditions and their parents face in shifting the management

responsibility to understand the challenges across conditions and the unique challenges specific to certain conditions, including those involving neurological sequelae or cognitive deficits. Further research should also assess the adolescent's readiness and their competency in taking on more management responsibility as well as develop and test interventions aimed at facilitating the shifting of management responsibility from the parent to the adolescent with chronic illness.

3.7 Conclusions

Shifting management responsibility from the parents to the adolescent with SCD is a critical and challenging process. Our findings indicate that the challenges that adolescents and parents face are adaptive-type challenges, rather than technical-type challenges. While technical-type challenges are easily fixed with technical work, more complex adaptive work is needed to address adaptive-type challenges (Anderson et. al, 2015), making the shifting of management responsibility more complex. However, collaboration and goal-setting across the adolescents- parent-provider triad will assist in facilitating the progressive shift of self-management responsibility from parents to adolescents and thus improve outcomes for adolescents with SCD and their parents

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4. Health Care Utilization by Transitioning Adolescents and Young Adults with Sickle Cell Disease

Sickle cell disease (SCD) is an inherited blood disorder affecting predominantly African Americans in the United States (Hassell, 2010). It is characterized by a variety of complications including vasoocclusive crises, acute chest syndrome, and stroke (Meier & Miller, 2012). With more than 95% of children with SCD surviving into adulthood (Lanzkron, Carroll, & Haywood, 2013; Quinn, Rogers, McCavit, & Buchanan, 2010), transitioning adolescents and young adults (AYAs) from pediatric to adult care is necessary to ensure developmentally appropriate health care services.

Health care transition can be defined as *“the purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from child-centered to adult-oriented health-care systems”* (Blum et al., 1993, p.570). The transition process should begin early in adolescence (American Academy of Pediatrics, et al., 2002), with transition readiness assessment and education on self-management, and independent life skills, among other skills (Cooley, 2013; Kauf, Coates, Huazhi, Mody-Patel, & Hartzema, 2009; McPheeters et al., 2014; Telfair, Myers, & Drezner, 1994; White, McManus, McAllister, & Cooley, 2012) and should continue throughout adolescence culminating in young adulthood with the transfer and integration into adult care.

Health care transition is a period of vulnerability for AYAs with SCD. For example, compared to younger children with SCD, AYAs are at higher risk for worse health outcomes including higher rates of complications such as vasoocclusive crises and strokes (Darbari et al., 2012; Lanzkron et al., 2013; Miller & Meier, 2012; Platt et al., 1994; Quinn et al., 2010); higher rates of affective conditions such as depression and anxiety (Benton, Ifeagwu, & Smith-Whitley, 2007; Jerrell, Tripathi, & McIntyre, 2011); and higher risk for mortality (Hamideh & Alvarez, 2013; Lanzkron et al., 2013). Despite the emphasis by a variety of federal agencies and professional organizations in the United States on the need for research focused on transition outcome measures that are sensitive to change and the identification of individual predictors for successful transition (McPheeters et al., 2014; Rosen, Blum, Britto, Sawyer, & Siegel, 2003), very few studies examined this in AYAs with SCD (Andemariam et al., 2014; Jordan, Swerdlow, & Coates, 2013; Newland, 2008; Telfair, Ehiri, Loosier, & Baskin, 2004; Telfair et al., 1994; Treadwell, Telfair, Gibson, Johnson, & Osunkwo, 2011). Further, studies that examined transition outcomes in AYAs with chronic illnesses focused on patient reported outcomes such as satisfaction with the process (McPheeters, et al., 2014). As a result, there is a limited understanding of the transition process in AYAs with SCD.

One important reason for exploring the transition process for AYAs with SCD is that they might be at risk for increased acute health care utilization due to the challenges

associated with unsuccessful transition. Some of these challenges include limited number of comprehensive sickle cell centers (Lotstein et al., 2006; Prabhakar et al., 2010; Smoth et al., 2006), limited number of adult providers who are adequately prepared and are comfortable to provide care for AYAs with SCD (McPeeters et al., 2014; Okumura et al., 2008), and maintaining health insurance once they transition to adult care (Lotstein et al., 2008). These challenges limit the AYAs' access to comprehensive SCD care and increase their reliance on the emergency department for their health care needs (Blinder et al., 2015). Further, the annual cost of health care for individuals with SCD is estimated to be as high as \$1.1 billion (Kauf et al., 2009) and decreasing health care costs associated with SCD necessitates an understanding of the patterns of health care utilization among individuals with SCD and the factors associated with these patterns.

However, there is a limited body of research exploring health care utilization in AYAs with SCD. Findings from these studies indicate that AYAs with SCD have higher rates of recurrent hospitalizations and emergency department encounters (Blinder et al., 2015; Brousseau, Owens, Mosso, Panepinto, & Steiner, 2010; Kauf et al., 2009; Wolfson et al., 2012). In particular, one study found that 18 to 30-year-old individuals with SCD had the highest hospitalization and re-hospitalization rates as well as the highest emergency department encounters compared to both younger and older individuals with SCD (Brousseau et al., 2010). Two other studies explored utilization across services in

individuals with SCD. Carroll and colleagues (2009) found a weak positive correlation between hospitalizations and clinic encounters, yet Epstein and colleagues (2006) found no association across these services.

Furthermore, there is a limited understanding of the factors that contribute to increased health care utilization in AYAs with SCD during health care transition. For example, it would be helpful to understand how individual characteristics, such as genotype and gender, are associated with patterns of health care utilization in AYAs with SCD. Currently, the state of knowledge on individual risk factors is inconclusive. While one study found that SCD severity was associated with a high risk for emergency department encounters (Aisiku et al., 2009) another found no association between SCD genotype and emergency department encounters in adult patients with SCD (Epstein et al., 2006). Similarly, while one study found a significant difference in health care utilization by gender, with females having less hospital and emergency department encounters and more clinic encounters (Epstein et al., 2006), another found no significant difference between males and females, but reported a tendency for males to have more encounters with higher percentage of hospital and emergency department stays than females (McClish et al., 2006). Other factors that might be associated with increased utilization include contextual factors such as the type of health insurance (Brousseau et

al., 2010; Schlenz, Born, Lackland, Adams, & Kanter, 2016) and location of residence (Schlenz et al., 2016; Telfair, Haque, Etienne, Tang, & Strasser, 2003; Wolfson et al., 2012).

In summary, there is limited research exploring health care utilization in AYAs, particularly during health care transition. Major limitations of the current studies include the use of cross-sectional designs and lack of longitudinal exploration of health care utilization trajectories. The pattern of co-utilization across services is also not clear. Finally, there is a limited understanding of the factors that might contribute to the increased health care utilization in the population AYAs with SCD who are transitioning to adult care. Exploring the trajectories of health care utilization across the transition period and the factors associated with health care utilization might assist in understanding this period of vulnerability and identify AYAs who are at risk of high utilization. Therefore, the purpose of this longitudinal database study was to address the current gap in the literature by identifying and describing the trajectories of health care utilization of transitioning AYAs with SCD and examining factors associated with these trajectories. Our overall goal was to identify and define successful transition for AYAs with SCD.

4.1 Theoretical Framework

Health care transition is a complex and multilevel phenomenon viewed from a developmental perspective by many pediatric, adolescent, and adult professional health

organizations (American Academy of Pediatrics, American Academy of Family Physicians, & American College of Physicians-American Society of Internal Medicine, 2002; Rosen, Blum, Britto, Sawyer, & Siegel, 2003). We used Bronfenbrenner's bioecological model of human development to explore health care transition as a complex developmental phenomenon across processes, person, context, and time (Bronfenbrenner, 2005; Bronfenbrenner & Morris, 2006; Griffin et al., 2013).

Processes are forms of regular, progressively more complex interactions between the individual and the environment that operate over time and are the primary mechanism producing human development (Bronfenbrenner, 2005; Bronfenbrenner & Morris, 2006). Processes in health care transition involve multiple reciprocal interactions between the AYA, their context, and the health care system and these interactions evolve over time. For example, these processes include the encounters that the AYA has with the health care system before, during, and after they transition from pediatric to adult care and how they utilize health care services to meet their health care needs. The magnitude of influence that these processes might have on health care transition would depend on the personal characteristics of the individual, the environmental context, and the period during which these processes take place (Bronfenbrenner, 2005; Bronfenbrenner & Morris, 2006). Individual characteristics, such as demographic characteristics and disease severity (Darling, 2007), might impact how the AYA interacts

with the health care system, the services they seek and receive, their health care outcomes, and as a result whether or not their health care transition is successful. Contextual factors (Bronfenbrenner & Ceci, 1994) such as health insurance status and location of residence have major implications for access to comprehensive SCD care for AYAs with SCD and might influence their health care utilization patterns. Finally, the timing of the actual transfer to adult care might have major implications on whether the AYA is ready for transition.

Using Bronfenbrenner's bioecological model of human development, this longitudinal database study had the following aims:

Aim one: Identify and describe the trajectories of health care utilization of transitioning AYAs (12 to 27 years old) with SCD, specifically

- 1) What are the classes of AYAs with SCD who had similar health care utilization trajectories during transition to adult care?
- 2) How does transfer to adult care occur?

Aim two: Examine the association between the utilization trajectory groups and the individual characteristics, contextual factors, and transfer related factors.

Specifically: What are the influences individual characteristics, contextual factors, and transfer related factors on the probability of class membership for health care utilization?

4.2 Methods

4.2.1 Databases and study population

Participants for this study were identified through the Duke Enterprise Data Unified Content Explorer (D.E.D.U.C.E.) research portal at Duke Health. D.E.D.U.C.E. research portal is a tool that provides access to clinical information collected as a by-product of patient care at Duke Health. Data sources include demographics, International Classification of Diseases diagnoses, procedures, medications, physician orders, lab data, among others. We extracted preexisting data from the D.E.D.U.C.E. research portal as well as the electronic medical records of AYAs with SCD who had encounters at Duke Health between January 1st, 1989 and July 30th, 2015. Data was extracted on participants who met the following inclusion criteria: AYA who 1) had SCD (Sickle cell anemia (SS); Sickle/Hb C disease (SC); Sickle/Beta plus thalassemia ($S\beta^+$) or Sickle/Beta zero thalassemia ($S\beta^0$) genotype), and 2) received at least two health care encounters with the pediatric sickle cell program at Duke Health during adolescence (12-18 years), with at least one encounter after age 15 years to ensure initial integration in pediatric services. AYAs who were carrier of the sickle cell trait (AS genotype) were excluded as they rarely have SCD complications, and therefore have a very different condition trajectory than AYAs with the other genotypes.

4.2.2 Measures

The selection of variables for the study was informed by Bronfenbrenner's bioecological model of human development. Table 3 presents the variables included in the study.

Table 3: Study variables

Theory construct	Study construct	Measures
Time	Age	<ul style="list-style-type: none"> • Age in years (12-27)
Individual characteristics	Sociodemographic Characteristics	<ul style="list-style-type: none"> • Age, gender, race, ethnicity, marital status, alive status (alive or deceased)
	Disease specific measures	<ul style="list-style-type: none"> • Genotype • Sickle cell disease complications (including vasoocclusive crises, acute chest syndrome) • Pediatric severity index for sickle cell disease-modified • Blood transfusions • Medications • Comorbidities
Context	Contextual factors	<ul style="list-style-type: none"> • Location of residence (distance to clinic) • Health insurance type
Proximal processes	Health care utilization	<ul style="list-style-type: none"> • Trajectories of sickle cell clinic visits (pediatric and adult) • Trajectories of hospitalizations and length of stay • Trajectories of emergency department encounters and length of stay • Age-related emergency department reliance score
	Transfer related factors	<ul style="list-style-type: none"> • Transfer age (First adult sickle cell clinic appointment) • Continuity of care: frequency of attending adult sickle cell clinic

Time. Time was defined in terms of participant's age from ages 12 to 27 years.

Individual characteristics. Individual characteristics included sociodemographic and disease specific measures. Sociodemographic variables included age, gender, race, ethnicity, marital status, and alive status (defined as alive or deceased) and were extracted using the D.E.D.U.C.E research portal. Disease specific measures included genotype, sickle cell disease complications, pediatric severity index for sickle cell disease (modified), blood transfusions, medications, and comorbidities. Genotype was defined as sickle cell anemia (SS), sickle/Hb C disease (SC); sickle/beta plus thalassemia ($S\beta^+$) or sickle/beta zero thalassemia ($S\beta^0$) and was extracted from provider notes in the electronic medical records.

Sickle cell disease related complications included vasoocclusive crises, acute chest syndrome episodes, presence of stroke or cerebral infarct, avascular necrosis, iron overload, chronic pain, lower extremity ulcers, priapism, among others. These variables were extracted through the D.E.D.U.C.E. research portal using ICD 9 codes and diagnoses names from encounter associated problem and diagnoses lists. We calculated age-related disease complications, namely the mean number of acute chest episodes and the mean number of vasoocclusive pain crises at ages 12 to 27 for the sample. We also calculated age-related sickle cell severity score at ages 15 and 19 using a modified version of the pediatric severity index developed by van den Tweel and colleagues

(2010). Age 15 was selected because it was one of our study inclusion criteria. Age 19 was selected because it was identified as the mean age at transfer to adult care for the sample (please see results section). In its original form, the severity index consists of 12 weighted items with a total score of 285 including lifetime cumulative incidence of avascular bone necrosis (AVN) of hip and/or shoulder, cerebral infarcts or vasculopathy, acute hepatic sequestration, pneumococcal meningitis/septicemia, priapism, and acute splenic sequestration; the cumulative incidence over a period of two years of acute chest syndrome (ACS) and vasoocclusive crisis (VOC); and Laboratory values of hemoglobin (g/dl), fetal hemoglobin (%), lactate dehydrogenase (LDH), and Leucocytes ($10^9/l$). The severity index had been shown in one study to differentiate between mild moderate, and severe disease (van den Tweel, van der Lee, Heijboer, Peters, & Fijnvandraat, 2010). Due to missing data in lab values for the sample, we modified the scale to include only weighted disease complications. We considered this to be acceptable because in the original validation study only seven (8%) participants out of 92 had scores on lab values (van den Tweel, van der Lee, Heijboer, Peters, & Fijnvandraat, 2010). We also excluded lifetime cumulative incidence of acute hepatic sequestration because there is no assigned ICD9 code for hepatic sequestration. We attempted to contact the authors of the original severity index to elicit their perspective on our modified version for the scale but were unsuccessful.

In addition, we extracted data on blood transfusions from diagnoses lists using ICD9 codes, procedure lists using CPT codes, and physician orders to capture all possible events related to blood transfusions in the database. We calculated the mean number of blood transfusions at ages 12 through 27 for the sample. If participants had more than six blood transfusions at age 15 or age 19 they were considered to be on chronic transfusions for that corresponding age.

The type of medications that the participants were receiving were extracted from outpatient medical reconciliation record through D.E.D.U.C.E. and medications were classified into hydroxyurea, extended release narcotics, short acting narcotics, and nonsteroidal anti-inflammatory classes.

Comorbidities examined included asthma and a variety of mental health conditions including anxiety, depression, adjustment disorders, conduct disorder, schizophrenia and psychotic disorders and were identified from encounter associated problem and diagnoses lists using ICD 9 codes and diagnoses names.

Contextual factors. Contextual factors included the location of residence (USPS 5-digit zip code) and the type of health insurance. Both variables were extracted using the D.E.D.U.C.E research portal. Distance in miles between the participant's location of residence and the sickle cell clinic was then calculated from participants' USPS 5-digit zip code and using the ZIPCITYDISTANCE function in SAS (version 9.4, SAS Institute,

Inc., Cary, NC). This function returns the geodetic distance in miles between two zip code locations. The type of health insurance was defined as private or public health insurance.

Proximal processes. Proximal processes included interactions between participants and the health care systems and were operationalized as health care utilization encounters and transfer related factors.

Health care utilization included encounters in the pediatric and adult sickle cell clinics, inpatient hospital encounters, and emergency department encounters over time. Emergency department encounters that resulted in hospitalizations were counted in both emergency department encounters and hospital encounters. We also calculated age-related emergency department reliance score for the sample at ages 12 to 27. Emergency department reliance score is defined as the proportion of emergency department visits to all ambulatory visits (clinic and emergency department encounters) and is used to differentiate between participants with high emergency department utilization due to increased care needs from participants with decreased access to ambulatory care (Kroner, Hoffmann, & Brousseau, 2010). We followed the defined cutoff point of 0.33 as set by the original authors for high (>0.33) and low (≤ 0.33) emergency reliance score (Kroner, Hoffmann, & Brousseau, 2010).

Transfer related variables included the age at transfer from pediatric to adult care defined as the age at which the participant had their first appointment in the adult sickle clinic. Continuity of care was examined as the frequency of adult clinic appointments after transfer to adult care. At least two encounters in the adult sickle cell clinic were needed for continuity and integration into adult care to be considered successful.

4.2.3 Procedures

After institutional review board approval, existing data on all AYAs with SCD who met the inclusion criteria was extracted using the cohort manager in the D.E.D.U.C.E. research portal. Filters using the inclusion criteria were sequentially applied to identify the study sample, following which data on study measures and variables was extracted from the portal and exported into SAS data files. A data technician with expertise in the D.E.D.U.C.E. portal assisted in the data extraction process to ensure the accuracy and rigor of the data set. Participants' electronic medical records were reviewed to extract additional variables not available in D.E.D.U.C.E (such as the patient's genotype). SAS statistical software (version 9.4, SAS Institute, Inc., Cary, NC) was used to manage and analyze the data with the exception of the Little's test for missing data (Little, 1988), which was conducted using SPSS statistical software (IBM SPSS Statistics 24).

4.2.4 Data analyses

Data analyses included computing descriptive statistics of sample characteristics. We corrected for outliers for continuous variables with the mean plus or minus four standard deviations.

For aim one, identify subgroups of transitioning AYAs with similar trajectories of health care utilization, we used latent class growth modeling (PROC TRAJ, version 9.4, SAS Institute, Inc., Cary, NC). Latent Class Growth Modeling identifies clusters of AYAs following a similar pattern of utilization (Jones & Nagin, 2007). First, we determined the number of latent trajectories classes (number of groups) that best describe the data by fitting models with different number of classes and comparing the fit indices of Bayesian information criteria (BIC) and significance ($p < 0.05$) to determine the most parsimonious model that was also useful and interpretable to the research aims. Next, we tested the adequacy of the selected model by examining the posterior probabilities of group membership and setting our limit to at least 0.7 for the posterior probability value, significance level at 0.05, and 95% confidence interval.

For aim two, examine the association between these trajectory groups and the individual characteristics, contextual factors, and transfer related factors, we used ordinal logistic regression. For this analysis, we conducted Little's missing completely at random (MCAR) test (Little, 1988) for all variables in our dataset. Once determined to be

missing at random, we used the impute function in SPSS to impute missing values then imported the new dataset into SAS and conducted the analysis. Group membership in a health care utilization trajectories identified in the latent class growth modeling was the outcome variable, and individual, contextual, and transfer related factors were the predictor variables. Due to the exploratory nature of the study we utilized stepwise selection as our model selection method for each of the health care services. Stepwise selection attempts to remove any non-significant variable from the model before adding a variable that is significant until the procedure is terminated when none of the variables meet the entry significance limit (SAS Institute Inc., 2016). Each addition and deletion of a variable is documented step by step in the output statement. Significance level was set at 0.1 to allow a variable into the model and 0.05 to allow a variable to remain in the model. Goodness of fit of the final selected model was determined by satisfaction of the model convergence criteria (GCON=1E-8), significant Likelihood Ratio test (beta=0) at $p \leq 0.05$, and improvement (decrease) of the Akaike Information Criteria (AIC) from the intercept only model to the intercept and covariate model.

4.3 Findings

4.3.1 Sample characteristics

Sociodemographic and contextual characteristics. The sample consisted of 339 participants born between 1977 and 1999, and the majority were still alive at their last

encounter (n=325, 95.87%). Fourteen patients passed away at a mean age of 24.6 years (SD=4.03). The majority of participants were Black (n=328, 96.76%), male (n=188, 55.46%), and single (n=292, 86.14%). Participants lived as close as 1.2 miles to as far as 282.4 miles from the clinic, with a mean of 48.55 miles. Of the 339 participants, 222 had complete information on their health insurance status and their insurance status changed between encounters over the study period, making it difficult to determine their overall insurance status. Table 4 presents the sample characteristics.

Table 4: Sample characteristics

N=339	n	%	Mean (SD)
Age Alive, in years	325	95.87	24.45 (5.38)
Age at death, in years	14	4.13	24.6 (4.03)
Gender			
<i>Male</i>	188	55.46	
<i>Female</i>	151	44.54	
Race			
<i>Black</i>	328	96.76	
<i>Other</i>	8	2.35	
<i>Unknown</i>	3	0.88	
Ethnicity			
<i>Other</i>	327	96.46	
<i>Hispanic</i>	2	0.59	
<i>Unknown</i>	4	1.18	
Marital Status			
Single	292	86.14	
<i>Married</i>	18	5.31	
<i>Unknown</i>	29	8.55	
Distance from clinic (in miles)	312	92	48.55 (32.56)

Disease specific measures. Participants' genotype, disease complications, and comorbidities are presented in table 5. Most participants had hemoglobin SS genotype

(n= 233, 68.73%), while 82 (24.19%) had Sickle/ Hb C disease (SC), 13 (3.83%) had Sickle/Beta plus thalassemia ($S\beta^+$), and 11 (3.21%) had Sickle/Beta zero thalassemia ($S\beta^0$). In terms of medical complications, most participants had a diagnosis of vasoocclusive crises (VOC) (n= 271, 79.94%), 138 (40.71%) had a diagnosis of acute chest syndrome episode (ACS), 76 (22.42%) had Cerebral infarcts or vasculopathy, and 15 (22.42%) had splenic sequestration. Of the 339 participants, 117 (34.51%) had a diagnosis of chronic pain.

We calculated the mean number of VOC and ACS episodes at ages 12 to 27 for the sample to create an annual rate of these two common complications with SCD. The mean number of VOC episodes ranged from five to 13 episodes at most ages except at age 12 and 16 where the mean number of VOC episodes was 21.33 and 30.44 respectively. However, these values had a wide 95% confidence interval that crossed zero indicating non-significance (figure 2). The mean number of ACS episodes seemed to be consistent across ages 12 to 27 with the mean ranging from one to four episodes (figure 3).

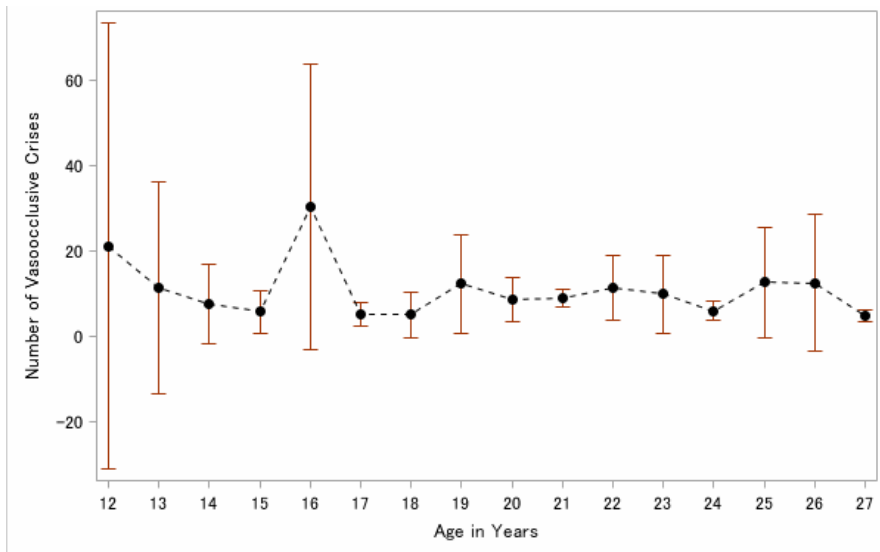


Figure 2: Mean number of vasoocclusive crises by age (ages 12-27) with 95% confidence interval

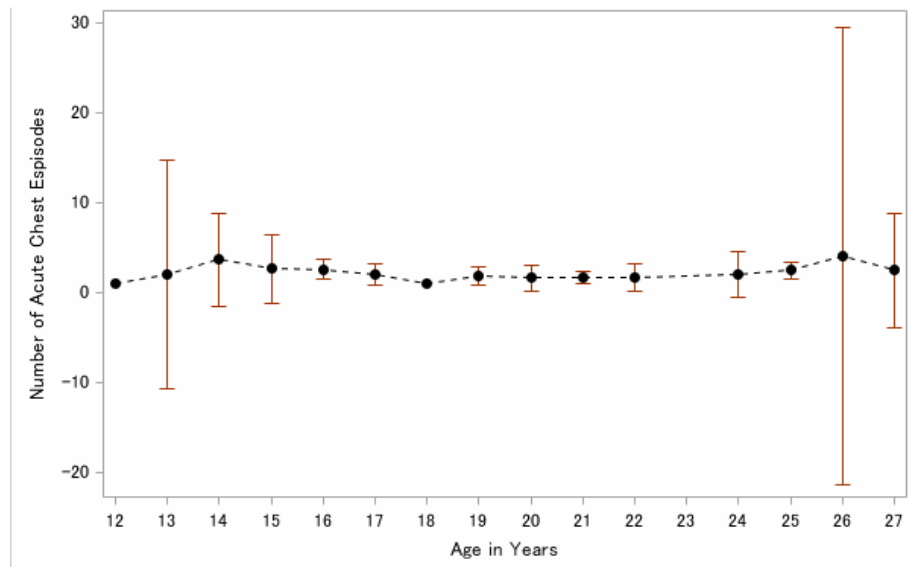


Figure 3: Mean number of acute chest syndrome episodes by age (ages 12-27) with 95% confidence interval

The mean modified sickle cell severity index at age 15 was 38.46 (SD= 24.66). It slightly decreased at age 19 to 35.83 (SD= 25.34). The distribution of the modified severity index at ages 15 and 19 are presented in figure 4.

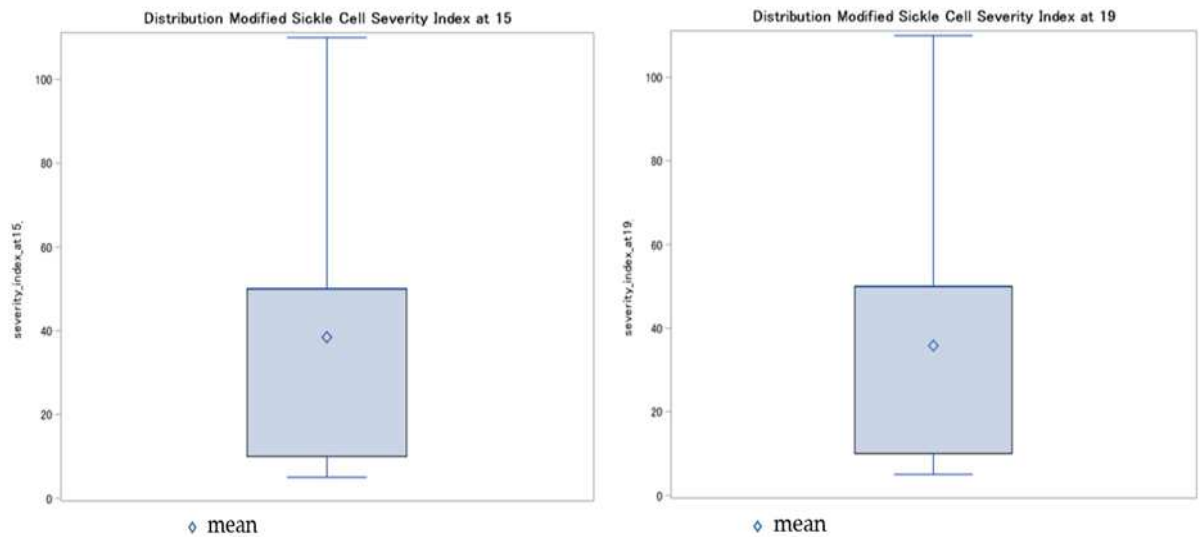


Figure 4: Mean modified sickle severity index at age 15 (left) & 19 (right)

Around 25% (n=88) of the sample had a diagnosis of asthma. While a variety of mental health conditions were prevalent in this sample, depression, anxiety, and adjustment disorders were the most common. Acute depression (n= 53, 15.63%) and major depression (n=25, 7.37%) episodes were prevalent alone and in combination with other mental health conditions (depression and anxiety n=17, 5.01%; adjustment disorder with depressive mood, n=17, 5.01%). A diagnosis of any depression episode

(including acute depression, major depression, depression with other conditions) was prevalent in 63 (18.58%) participants. Seven participants had a diagnosis of suicidal ideation (2%) and four (11.8%) attempted suicide. None were successful and were all alive at the last encounter in the dataset. The age at which participants attempted suicide was as young as 16 and as old as 32. One participant attempted suicide four times between the ages of 23 and 24. When we examined their characteristics, all four participants were black and had SS genotype. Three were male and one was female. Their modified severity index at ages 15 and 19 ranged between 50 and 80. All four participants had diagnoses of cerebral infarcts/ vasculopathy, chronic pain, major depression, and anxiety. Only one participant was diagnosed with suicidal ideation.

Forty-eight (14.16%) participants had a diagnosis of anxiety and 12 (3.88%) had attention deficit disorder or attention deficit hyperactivity disorder. We also examined the prevalence of physical and mental disabilities in the sample. One participant was legally blind, one was hearing impaired, and seven had mild to severe intellectual disabilities.

We calculated the rate of blood transfusions at ages 12 to 27 for the 86 (25.35%) participants who received blood transfusions. Overall, participants who received blood transfusions had between 18 and 80 blood transfusions a year from ages 12 to 27 years. High rates of blood transfusions occurred at ages 16 (mean= 33.88; 95% CI= 9.72-57.05),

age 18 (mean= 25.54; 95% CI= 5.72-45.35), age 19 (mean= 28.23; 95%CI= 15.81-40.65), age 20 (mean= 25.88; 95%CI= 9.73-42.02), age 21 (mean= 31.27; 95%CI= 13.94-48.6), and age 25 (mean= 50.3; 95%CI= 24.01-76.5). High rates of blood transfusions were found at age 22, 23, 26, and 27, but these ages had a 95% confidence interval crossed zero (figure 5).

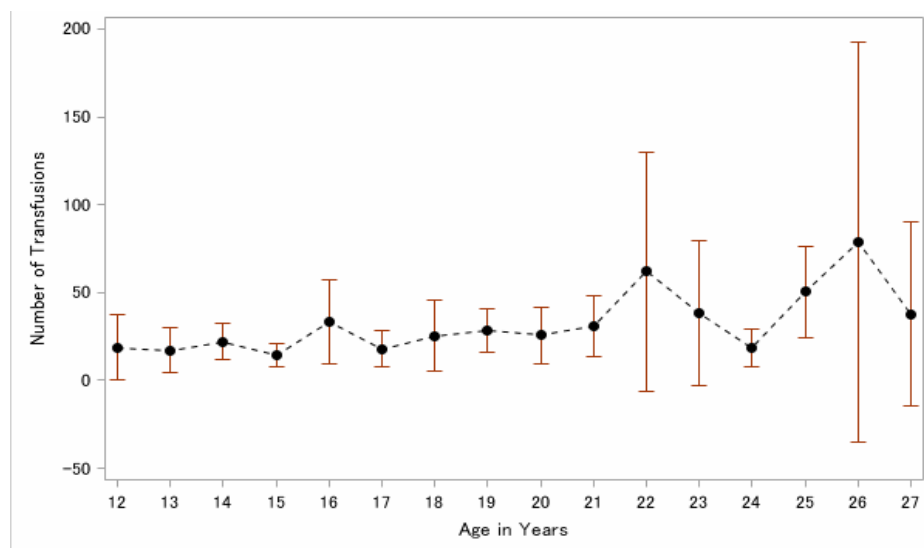


Figure 5: Mean number of blood transfusions by age (ages 12-27) with 95% confidence interval

Of the 339 patients, 248 (73.16%) had data on their outpatient medications. The majority of these participants were on analgesics/analgesic combinations/ non-steroidal anti-inflammatory (n=185, 54.58%), 168 (49.56%) were on short acting narcotics, and 35 (10.32%) were on long acting narcotics for pain management. Around 50% (n=168) were on hydroxyurea.

Table 5: Participants' disease complications, comorbidities, and outpatient medications

N=339	n	%	Mean (SD)
Genotype	339	100	
<i>Sickle cell anemia (SS)</i>	233	68.73	
<i>Sickle/Hb C disease (SC)</i>	82	24.19	
<i>Sickle/Beta plus thalassemia (Sβ+)</i>	13	3.83	
<i>or Sickle/Beta zero thalassemia (Sβ°)</i>	11	3.24	
Sickle cell complications			
<i>Severity Score at age 15</i>	153	45.13	38.46(24.66)
<i>Severity Score at age 19</i>	174	51.33	35.83(25.34)
<i>Vas-occlusive pain crisis</i>	271	79.94	
<i>Acute chest syndrome</i>	138	40.71	
<i>Avascular necrosis of hip/ shoulder</i>	21	6.19	
<i>Avascular necrosis, other</i>	75	22.12	
<i>Splenic sequestration</i>	15	4.42	
<i>Cerebral infarcts/ vasculopathy</i>	76	22.42	
<i>Seizures</i>	25	7.37	
<i>Pneumococcal meningitis/ septicemia</i>	2	0.59	
<i>Priapism</i>	37	10.91	
<i>Pulmonary hypertension</i>	34	10.03	
<i>Iron-overload, transfusion related</i>	28	8.26	
<i>Jaundice</i>	38	11.21	
<i>Pain, chronic</i>	117	34.51	
<i>Ulcer, lower extremity</i>	6	1.77	
<i>Ulcer, lower extremity, chronic</i>	1	0.29	
Medical Comorbidities			
<i>Asthma</i>	88	25.96	
Mental health comorbidities			
<i>Depression, any</i>	63	18.58	
<i>Depression, acute</i>	53	15.63	
<i>Depression, major</i>	25	7.37	
<i>Depression and anxiety</i>	17	5.01	
<i>Suicidal ideation</i>	7	2.06	
<i>Suicide</i>	4	1.18	
<i>Anxiety</i>	48	14.16	
<i>Attention deficit /Attention deficit hyperactivity disorder</i>	12	3.54	
<i>Adjustment disorder with anxiety & depression</i>	3	0.88	
<i>Adjustment disorder with anxiety</i>	2	0.59	

<i>Adjustment disorder with depression</i>	17	5.01
<i>Adjustment disorder, other</i>	6	1.77
<i>Mood disorder, other</i>	3	0.88
<i>PTSD</i>	5	1.47
<i>Paranoid Schizophrenia</i>	2	0.59
<i>Psychotic disorder</i>	7	2.06
<i>Schizoaffective disorder</i>	2	0.59
<i>Conduct disorder</i>	1	0.29
<i>Impulse control</i>	1	0.29
Learning problems	1	0.29
Disabilities		
<i>Legally blind</i>	1	0.29
<i>Hearing impaired</i>	1	0.29
<i>Intellectual disabilities</i>	7	2.06
Blood transfusions	86	25.37
Outpatient Medications	248	73.16
<i>Analgesics/Analgesic combinations/ NSAIDS</i>	185	54.58
<i>Narcotics, short acting</i>	167	49.26
<i>Narcotics, long acting</i>	35	10.32
<i>Hydroxyurea</i>	168	49.56
<i>Medications Unknown</i>	91	26.84

4.3.2 Health Care Utilization

Table 6 presents a description of the total health care encounters over the study period by service type. The 339 participants had a total of 10,848 pediatric and adult sickle cell clinic encounters. Of the 339 participants, 240 (70.8%) had a total of 3,840 hospital encounters and 197 (58.1%) had a total of 3,152 emergency department (ED) encounters. The mean length of stay for hospitalizations was five days (SD=5.5, median=3.55), while the mean length of stay for ED encounters was 7.64 hours (SD=5.3, median=6.48). The distribution of the mean length of stay in days for hospitalizations

and the distribution of the mean length of stay in hours for the ED are presented in figures 6 and 7 respectively.

Table 6: Health care encounters by service type over the study period

Service	n (%)	No. of Encounters
Sickle Cell Pediatric and Adult Clinics	339 (100)	10,848
Hospital	240 (70.8)	3,840
Emergency Department	197 (58.1)	3,152

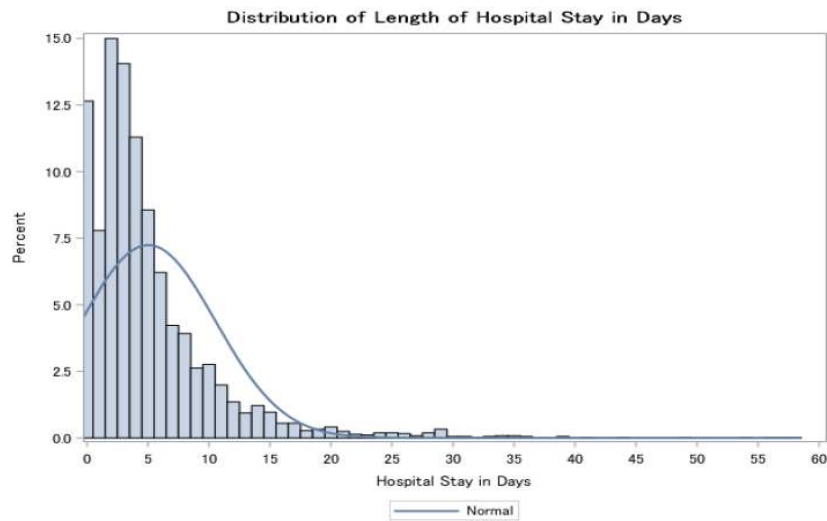


Figure 6: Mean length of stay in days for all hospitalizations

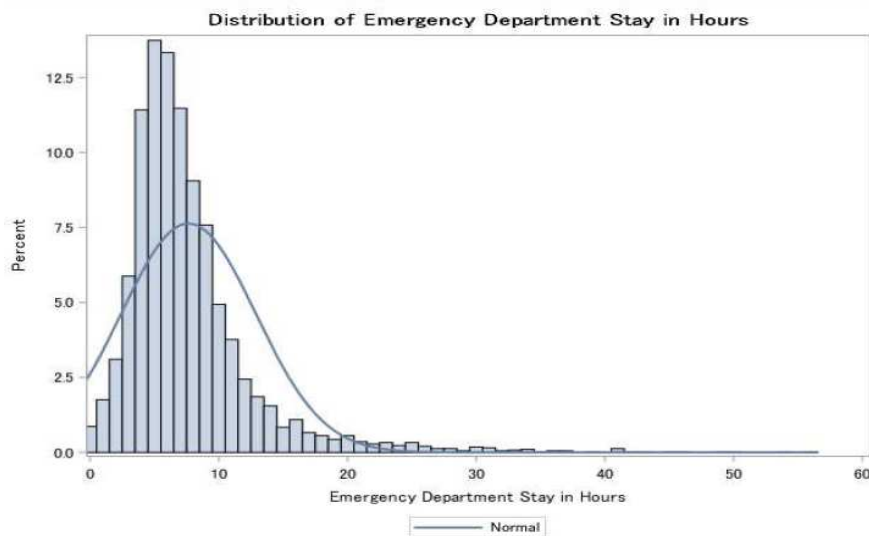


Figure 7: Mean length of stay in hours for all emergency department encounters

To identify the trajectories of health care utilization for transitioning AYAs with SCD (aim one) we conducted a latent class growth modeling. Next, we present the results of the latent class growth modeling by service type.

4.3.2.1 Pediatric and Adult Clinic Utilization Trajectories

A three-group model resulted in the best fit model for the data for clinic encounters based on Bayesian Information Criterion (BIC), significance ($p \leq 0.005$), 95% confidence intervals, group membership probabilities, and interpretability. Figure 8 shows the three trajectory groups identified in the model. The majority of participants ($n=206$, 60.77%) were in group one characterized by a low utilization trajectory across both pediatric and adult sickle cell clinics. A plateau is observed in group one between the ages of 13 and 16 which was due to an equal number of visits at ages 14 and 15.

Group two included 28.02% (n=95) of participants who had a medium pediatric and medium adult clinic utilization trajectory. This group appeared to have had a decreasing number of encounters in the adult clinic after age 23. Finally, a third group of 38 (11.21%) participants had a trajectory of high pediatric- high adult clinic utilization. This group also appears to have had a decreasing number of encounters in the adult clinic after age 23, but seems to increase slightly at ages 26 to 27.

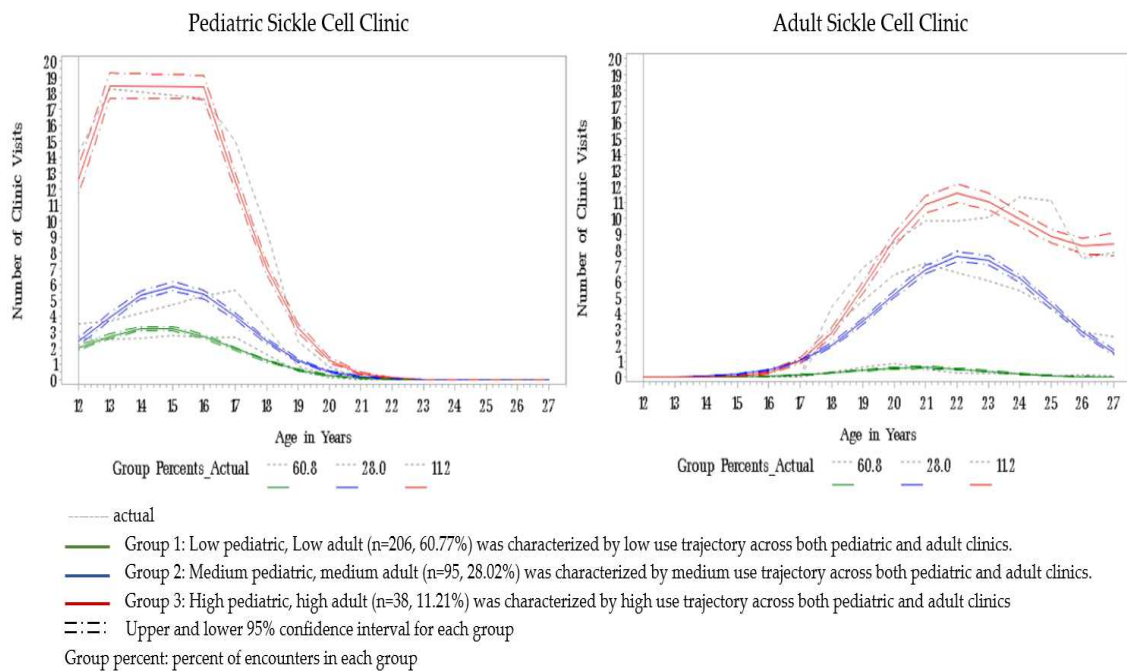


Figure 8: Pediatric (left) and adult (right) sickle cell clinic utilization trajectories by participant age (ages 12-27)

4.3.2.2 Hospital Utilization Trajectories

The three-group model was also the best fit model for the hospital encounter data based on Bayesian Information Criterion (BIC), significance ($p \leq .005$), 95% confidence intervals, group membership probabilities, and interpretability. Figure 9 shows the three trajectory groups identified in the model. Patterns included trajectories of low hospital utilization group with the majority of participants ($n=171$, 71.25%) in this group, a medium hospital utilization group ($n= 54$, 22.5%), and a high hospital utilization group ($n=15$, 6.25 %) with increasing number of encounters with increasing age.

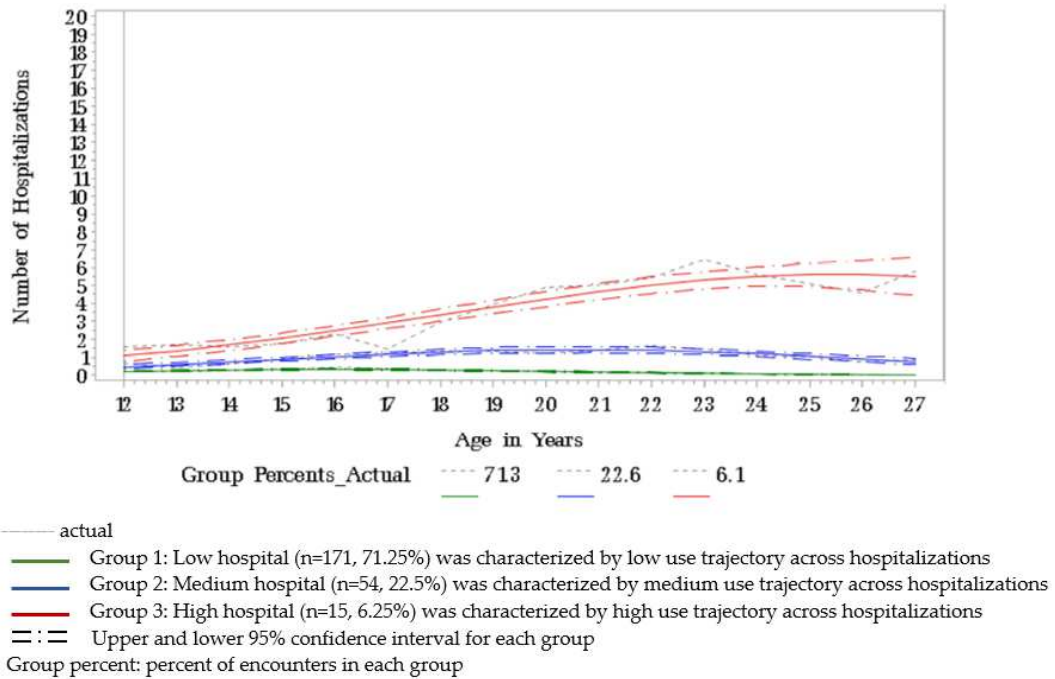


Figure 9: Hospital utilization trajectories by participant age (ages 12-27)

4.3.2.3 Emergency Department Utilization Trajectories

The three-group model was the best fit for the emergency department encounter data based on Bayesian Information Criterion (BIC), significance ($p \leq .005$), 95% confidence interval, group membership probabilities, and interpretability. Figure 10 shows the three trajectory groups identified in the model. The majority of participants had a low ED utilization trajectory ($n=146$, 74.11%). A second group of participants ($n=39$, 19.8%) had a medium ED utilization trajectory and a small group of participants ($n=12$, 6.09%) had a high ED utilization trajectory. Starting age 17 years, the high utilization group steadily increased in the number of ED encounters.

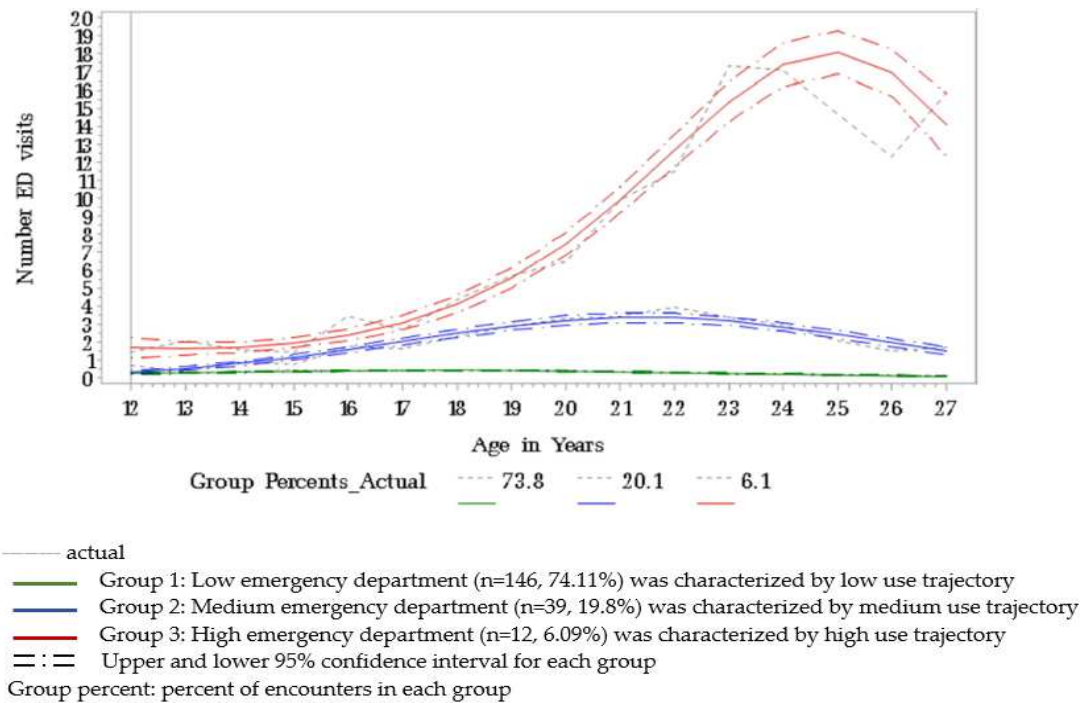


Figure 10: Emergency department utilization trajectories by participant age (ages 12-27)

4.3.2.3 Coutilization Across Health Services

We examined the hospital and ED coutilization for each of the clinic trajectory groups to explore participant utilization across services.

Low pediatric, low adult clinic utilization group (n=206). Overall hospital coutilization for this group occurred as follows: 84 (40.78%) participants had no hospital, 116 (56.31%) had low hospital, and only six (2.91%) had medium hospital coutilization. Overall ED coutilization for this group occurred as follows: 120 (58.25%) had no ED, 83 (40.29%) had low ED, and only three (1.46%) participants had medium ED coutilization. Other coutilization patterns were found in which around 34% (n=70) of the low pediatric low adult clinic utilizer typology had no hospital or ED coutilization and only three participants had medium ED and low or medium hospital coutilization trajectories. Overall, the majority of participants who had low pediatric and low adult clinic utilization trajectories had no to low hospital and ED coutilization trajectories.

Medium pediatric, medium adult clinic utilization group (n=95). Overall hospital coutilization for the medium pediatric, medium adult clinic utilization group occurred as follows: the majority had low hospital (n=39, 41.05%) or medium hospital (n=37, 38.95%) utilizations. Only 11 (11.58%) participants had no hospital and another eight (8.42%) had high hospital coutilization. Overall ED coutilization for this group occurred as follows: the majority (n=48, 50.53%) had low ED utilization followed by

medium ED utilization for 24 (25.26%) participants. Only 15 (15.79%) participants had no ED and eight (8.42%) had high ED utilization. The majority (n=40, 42.11%) in this group also had low hospital and low or no ED utilization. Finally, only six (6.32%) participants had high hospital and high ED utilization and another 4 (4.22%) participants had medium to high ED and medium to high hospital utilization.

High pediatric, high adult utilization group (n=38). Overall hospital utilization for this group occurred as follows: 16 (42.11%) had low hospital and 11 (28.95%) had medium hospital utilization. Only four (10.53%) participants had no hospital and another seven (18.42%) had high hospital utilization. Overall ED utilization for this group was as follows: 15 (39.47%) participants had low ED followed by 12 (31.58%) participants who had medium ED utilization. Seven (18.42%) participants had no ED and four (10.53%) participants had high ED utilization. The four participants with high ED utilization also had high hospital utilization. Around 34.6% had medium or high hospital and another 10 had medium ED and medium to high hospital utilization. Only three patients did not have any ED or hospital utilization in this group.

4.3.3.4 Emergency department reliance

To differentiate between participants with high emergency department utilization due to increased care needs from participants with decreased access to

ambulatory care (Kroner, Hoffmann, & Brousseau, 2010), we calculated age-related emergency department reliance scores (EDR) for participants who had emergency department encounters (n= 197, 58.1%). The distribution of the mean EDR score at age 12 to age 27 are presented in figure 10. The lowest mean EDR was 0.035 at age 12 and highest at age 27 with a mean of 0.151. The mean EDR score steadily increased by age, particularly after age 18, indicating an increased risk for high EDR scores with increased age. Although the mean EDR scores at ages 12 to 27 were still lower than the conventional cutoff point of 0.33 for high emergency department reliance (Kroner, Hoffmann, & Brousseau, 2010), the distribution of these means indicated that some participants had EDR scores that were as high as 0.667-0.96. We ran a secondary analysis of the frequency of participants with high (≥ 0.33) EDR score at each age. The results are shown in figure 11. There was a trend of increasing percentage of participants with high EDR scores associated with increased age.

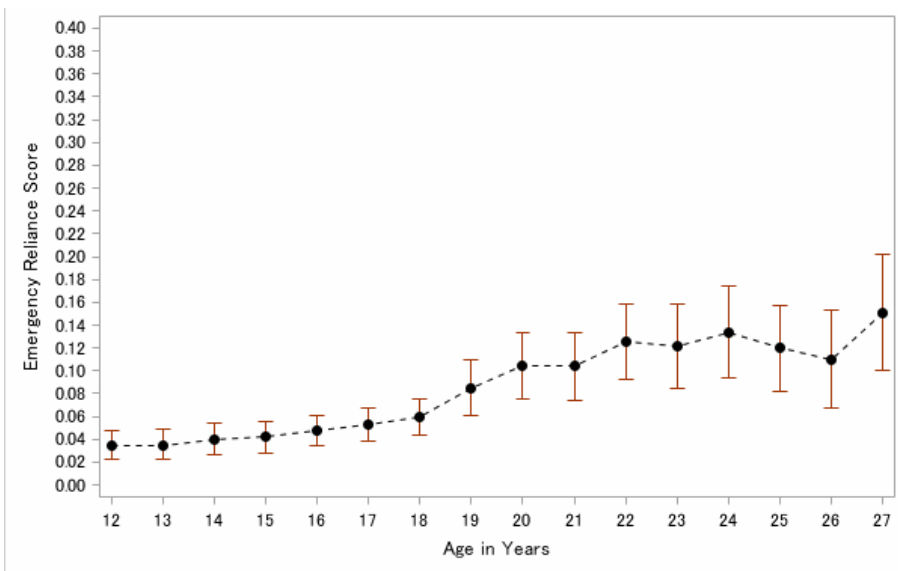


Figure 11: Mean emergency department reliance score by age (ages 12-27) with 95% confidence interval

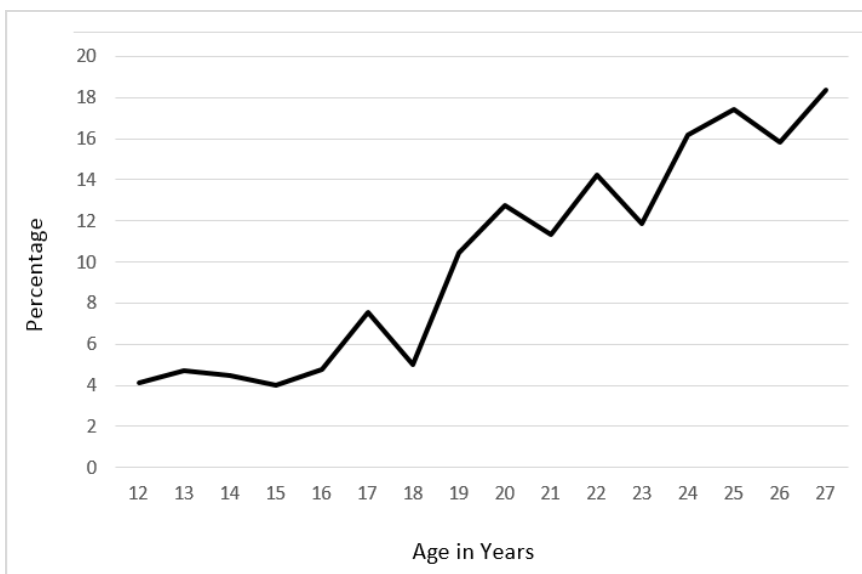


Figure 12: Percentage of participants with high emergency department reliance score (>0.33) by age

4.3.4 Transfer to Adult Care and Continuity of Care

The age of transfer to adult care was defined as the age at first encounter in the adult sickle cell clinic. Of the 339 patients in the sample, 223 had a first encounter in the adult sickle cell clinic. The mean age at transfer was 18.76 (SD=1.54). Figure 13 shows the distribution of the mean age at transfer to the adult sickle cell clinic.

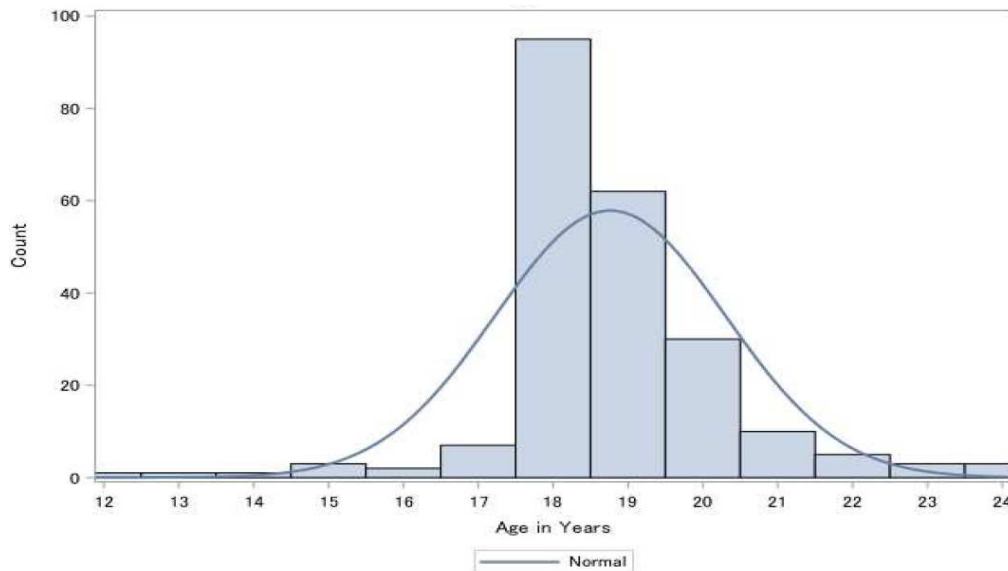


Figure 13: Mean age at transfer to adult sickle cell clinic

We also examined the continuity of care after transfer. Of the 223 participants who transferred to adult care, 210 (94.17%) had at least one additional encounter in the adult sickle cell clinic, while 13 (5.83%) had only one adult encounter (at transfer). The range of encounters in the adult sickle cell clinic was widely distributed from 1 to 220 encounters after transfer with 65% of the participants who transferred having more than

ten encounters at the adult sickle cell clinic and around 10% having more than 100 encounters indicating appropriate integration into adult care. Figure 14 shows the distribution of the number of adult sickle cell clinic encounters after transfer to adult care.

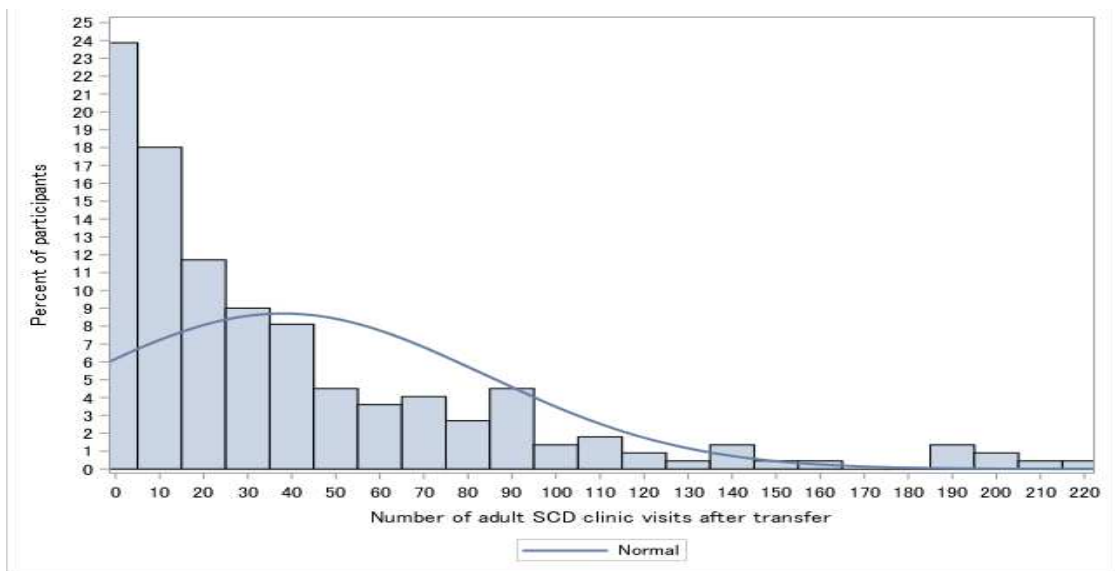


Figure 14: Continuity of care after transfer to adult care

4.3.5 Factors Associated with Health Care Utilization Trajectory Groups

Characteristics associated with group membership in each of the service utilization trajectories including individual characteristics, contextual factors, and transfer related factors were examined. Little's test indicated that data was missing at random (chi square=593.898, df=726, p=1.000). We used the imputation method in SPSS

to impute missing values and imported the new dataset into SAS to conduct the ordinal logistic regression procedure. The fit indices for the ordinal logistic models selected by the stepwise selection procedure are presented in table 7.

Table 7: Fit indices for the regression models selected for the sickle cell clinic, hospital, and emergency department

Model	Convergence criterion GCONV=1E-8	Model fit statistics (AIC)		Likelihood Ratio Test		
		Intercept only	Intercept & covariates	Chi-square	Df	p
Clinic	satisfied	617.250	371.933	263.3164	9	<0.0001
Hospital	satisfied	617.250	534.17	95.0799	6	<0.0001
Emergency Department	satisfied	284.974	124.574	178.3998	9	<0.0001

4.3.5.1 Factors associated with clinic trajectory groups

The clinic model selected satisfied the convergence criterion (GCONV=1E-8) and Likelihood Ratio test (chi-square=263.3164, df=9, p<0.0001). AIC also improved in the model with the intercepts and the predictor variables (AIC=371.933) compared to the intercepts only model (AIC= 617.250). The results of the ordered log odds (logit) regression coefficients are presented in table 8. The estimated log odds of high vs. medium and low clinic utilization when all predictor variables were estimated at zero was -10.5474 (p<0.0001), while the estimated log odds of high and medium vs. and low

clinic utilization when the predictor variables were estimated at zero was -7.1943 (p<0.0001).

Participants' age (p<0.0001) and their location of residence (distance from clinic, p<0.0001) were significant individual and contextual characteristics that predicted clinic group membership. Cerebral infarction (p<0.0001), iron overload (p<0.0001), the number of blood transfusions at age 19 (p=.0113), lower extremity ulcer (p=0.0174), and being on hydroxyurea (p=0.0007) were significant sickle cell complications that predicted clinic group membership. Attention deficit disorders (p=0.0003) were a significant comorbidity that predicted of sickle cell clinic utilization trajectories. Finally, age at the time of transfer to adult care (p<0.0001) was a significant transfer related factor that predicted clinic group membership. Table 8 lists the parameters and their corresponding Wald test, while table 9 lists the odds ratios for each of the significant predictors with their corresponding 95% confidence interval.

Individual characteristics.

Age. Age was the only significant sociodemographic characteristic for predicting clinic group membership (OR=1.2; 95% CI, 1.125-1.28). For every one-year increase in participant's age, the odds of high clinic utilization group were 1.2 times greater than for the combined medium and low clinic utilization groups, given that all other variables

were held constant. Likewise, for every one-year increase in age, the odds of middle and low utilization groups were 1.2 times greater than for the high utilization group.

Disease specific measures. Iron overload had the highest odds ratio in the model (OR=36.611; 95% CI=11.657-114.979). As participant attained this complication, the odds of high clinic utilization were 36.611 times greater than for the combined medium and low clinic utilization groups, given that all other predictors are held constant. Similarly, as participants acquired iron overload, the odds of middle and high vs. low clinic utilization was 36.611 times greater, given that all other predictors are held constant.

The next highest odds ratio in the model was that of lower extremity ulcers (OR=8.528; 95%CI, 1.45-49.906). As participant developed lower extremity ulcer, the odds of high clinic utilization were 8.528 times greater than for the combined medium and low clinic utilization groups, given that all other predictors were held constant. Similarly, as participants went from no lower extremity ulcer to having a lower extremity ulcer, the odds of middle and high vs. low clinic utilization were 8.528 times greater, given that all other predictors were held constant.

Another sickle cell complication that was predictive of clinic group membership was Cerebral infarct/ vasculopathy (OR=7.226; 95%CI, 3.599-14.510). As participant developed a cerebral infarct/ vasculopathy, the odds of high clinic utilization were 7.226 times greater than for the combined medium and low clinic utilization groups, given

that all other predictors were held constant. Similarly, as participants went from no cerebral infarct/ vasculopathy to the presence of a cerebral infarct/ vasculopathy, the odds of middle and high vs. low clinic utilization was 7.226 times greater, given that all other predictors were held constant.

While being on chronic transfusion was not a significant predictor for clinic group membership, the total number of blood transfusions at age 19 was a significant predictor (OR=1.053; 95%CI, 1.012-1.097). For every one-count increase in the total number of transfusions at age 19, the odds of high clinic utilization group were 1.053 times greater than for the combined medium and low clinic utilization groups, given that all other variables were held constant. Likewise, for every one-count increase in the total number of transfusions at age 19, the odds of middle and low utilization groups were 1.053 times greater than for the high utilization group.

The only medication that was predictive of clinic group membership was hydroxyurea (OR=3.175; 95%CI, 1.626-6.198). As participant went from not receiving hydroxyurea to receiving hydroxyurea, the odds of high clinic utilization were 3.175 times greater than for the combined medium and low clinic utilization groups, given that all other predictors were held constant. Similarly, as participants were placed on hydroxyurea, the odds of middle and high vs. low clinic utilization was 3.175 times greater, given that all other predictors were held constant.

The only comorbidity that was significant for clinic group membership was attention deficit disorders (attention deficit disorder and attention deficit disorder with hyperactivity, OR=11.487; 95%CI, 3.033-43.505). As participant were diagnosed with attention deficit disorders, the odds of high clinic utilization were 11.487 times greater than for the combined medium and low clinic utilization groups, given that all other predictors were held constant. Similarly, as participants were diagnosed with attention deficit disorders, the odds of middle and high vs. low clinic utilization was 11.487 times greater, given that all other predictors were held constant.

Contextual factors. While we could not examine the effect of type of health insurance on clinic group membership, distance between the participant's location of residence and the sickle cell clinic was a significant predictor of clinic group membership (OR=0.980; 95% CI, 0.970-0.989). For every one-mile increase in distance between the home and the clinic, the odds of high clinic utilization group were 0.98 times less than for the combined medium and low clinic utilization groups, given that all other variables were held constant. Likewise, for every one-mile increase in distance, the odds of middle and low clinic utilization groups are 0.98 times lower than for the high clinic utilization group.

Transfer related factors. Participants' age at transfer to adult care was a significant predictor of clinic group membership (OR=1.115; 95%CI, 1.061-1.172). For

every one-year increase in participant age at transfer, the odds of high clinic utilization group were 1.115 times more than for the combined medium and low clinic utilization groups, given that all other variables were held constant. Likewise, for every one-year increase in age at transfer, the odds of middle and low utilization groups are 1.115 times greater than for the high utilization group, given that all other variables were held constant.

Table 8: Parameter estimates and corresponding Wald test for clinic model

Analysis of Maximum Likelihood Estimates						
Parameter		Df	Estimate	Standard Error	Wald Chi-Square	Pr > ChiSq
Intercept	High clinic	1	-10.5474	1.0895	93.7175	<0.0001
Intercept	Medium clinic	1	-7.1943	0.9354	59.1523	<0.0001
Cerebral Infarct		1	1.9777	0.3557	30.9187	<0.0001
Age		1	0.1822	0.0329	30.6019	<0.0001
Iron Overload		1	3.6003	0.5839	38.0212	<0.0001
Age at first adult		1	0.1086	0.0254	18.2347	<0.0001
Attention deficit disorders		1	2.4412	0.6794	12.9096	0.0003
Total transfusions at 19		1	0.0520	0.0205	6.4119	0.0113
Distance		1	-0.0207	0.00500	17.0596	<0.0001
Hydroxyurea		1	1.1552	0.3413	11.4536	0.0007
Lower Extremity Ulcer		1	2.1434	0.9014	5.6538	0.0174

Table 9: Odds ratio estimates and corresponding 95% confidence interval

Effect	Odds Ratio Estimates		
	Point Estimate	95% Wald Confidence Limits	
Cerebral Infarct	7.226	3.599	14.510
Age	1.200	1.125	1.280
Iron Overload	36.611	11.657	114.979
Age at first adult	1.115	1.061	1.172
Attention deficit disorders	11.487	3.033	43.505
Total transfusions at 19	1.053	1.012	1.097
Distance	0.980	0.970	0.989
Hydroxyurea	3.175	1.626	6.198
Lower Extremity Ulcer	8.528	1.457	49.906

4.3.5.2 Factors associated with hospital trajectory groups

The hospital model selected satisfied the convergence criterion (GCONV=1E-8) and Likelihood Ratio test (chi-square=95.0799, df=6, $p<0.0001$). AIC also improved in the model with the intercepts and the predictor variables (AIC=534.17) compared to the intercepts only model (AIC= 617.250). The results of the ordered log odds (logit) regression coefficients are presented in table 10. The estimated log odds of high vs. medium and low hospital utilization when all predictor variables were estimated at zero was -3.8070 ($p<0.0001$) while the estimated log odds of high and medium vs. and low

hospital utilization when the predictor variables were estimated at zero was -1.6439 (p<0.0001).

No significant contextual or transfer related factors predicted hospital group membership. Only individual characteristics were significant. Chronic transfusion (defined as more than six annual transfusions) at age 19 (p=0.0184), diagnosis of chronic pain (p<0.0001), and being on hydroxyurea (p=0.0343) and long-acting narcotics (p=0.0254) were significant sickle cell complications that predicted hospital group membership. Being diagnosed with depression (p=0.018) and having intellectual disabilities (p=0.0129) were significant comorbidities that predicted hospital utilization trajectory group. Table 10 lists the parameters and their corresponding Wald test, while table 11 lists the odds ratios for each of the significant predictors with their corresponding 95% confidence interval.

Individual characteristics

Disease specific measures. Being diagnosed with chronic pain also was a significant predictor of hospital group membership (OR=3.253; 95%CI, 1.916-5.522). When participants had a chronic pain diagnoses, the odds of high hospital utilization were 3.253 times greater than for the combined medium and low hospital utilization groups, given that all other predictors were held constant. Similarly, as participants moved from no diagnosis of chronic pain to a diagnosis of chronic pain, the odds of

middle and high vs. low hospital utilization were 3.253 times greater, given that all other predictors were held constant.

The odds ratio for chronic blood transfusion at age 19 were 3.923 (95%CI, 1.259-12.22). As participants moved from not receiving chronic transfusions to receiving chronic transfusions at age 19, the odds of high hospital utilization were 3.923 times greater than for the combined medium and low hospital utilization groups, given that all other predictors were held constant. Similarly, as participants moved from not receiving chronic transfusions to receiving chronic transfusions at age 19, the odds of middle and high vs. low hospital utilization were 3.923 times greater, given that all other predictors were held constant.

Use of long-acting narcotics (OR=2.34; 95%CI, 1.11-4.933) and hydroxyurea (OR=1.871; 95%CI, 1.047-3.341) were predictive of hospital group membership. As participants moved from not being on long-acting narcotics to being on long-acting narcotics, the odds of high hospital utilization were 2.34 times greater than for the combined medium and low hospital utilization groups, given that all other predictors were held constant. Similarly, as participants went from not being on long-acting narcotics to being on long-acting narcotics, the odds of middle and high vs. low hospital utilization were 2.34 times greater, given that all other predictors were held constant.

For hydroxyurea, as participants moved from not being on hydroxyurea to being on hydroxyurea, the odds of high hospital utilization were 1.871 times greater than for the combined medium and low hospital utilization groups, given that all other predictors were held constant. Similarly, as participants went from not being on long-acting narcotics to being on long-acting narcotics, the odds of middle and high vs. low hospital utilization were 1.871 times greater, given that all other predictors were held constant.

Two comorbidities were significant predictors for hospital group membership: intellectual disabilities (OR=7.96; 95%CI, 1.551-40.858) and diagnosis of depression (OR=2.212; 95%CI=1.146-4.271). Intellectual disabilities (mild to severe) had the highest odds ratio. As participants were diagnosed with an intellectual disability, the odds of high hospital utilization were 7.96 times greater than for the combined medium and low hospital utilization groups, given that all other predictors were held constant. Similarly, as participants were diagnosed with an intellectual disability, the odds of middle and high vs. low hospital utilization were 7.96 times greater, given that all other predictors were held constant.

Depression was another comorbidity that predicted hospital group membership. As participants were diagnosed with depression, the odds of high hospital utilization were 2.212 times greater than for the combined medium and low hospital utilization

groups, given that all other predictors were held constant. Similarly, as participants were diagnosed with any form of depression, the odds of middle and high vs. low hospital utilization were 2.212 times greater, given that all other predictors were held constant.

Table 10: Parameter estimates and their corresponding Wald test for hospital model

Analysis of Maximum Likelihood Estimates

Parameter		DF	Estimate	Standard Error	Wald Chi-Square	Pr > Chi Sq
Intercept	High hospital	1	-3.7449	0.3226	134.7737	<0.0001
Intercept	Medium hospital	1	-1.6381	0.2394	46.8159	<0.0001
Depression		1	0.7941	0.3356	5.6000	0.0180
Chronic pain		1	1.1795	0.2701	19.0761	<0.0001
Narcotics, Long-acting		1	0.8502	0.3805	4.9931	0.0254
Intellectual disabilities		1	2.0744	0.8346	6.1785	0.0129
Hydroxyurea		1	0.6262	0.2959	4.4775	0.0343
Chronic transfusion at age 19		1	1.3669	0.5797	5.5599	0.0184

Table 11: Odds Ratio Estimates and corresponding 95% confidence interval

Odds Ratio Estimates			
Effect	Point Estimate	95% Wald Confidence Limits	
Depression	2.212	1.146	4.271
Chronic pain	3.253	1.916	5.522
Narcotics, Long-acting	2.340	1.110	4.933
Intellectual disabilities	7.960	1.551	40.858
Hydroxyurea	1.871	1.047	3.341
Chronic transfusion at age 19	3.923	1.259	12.220

4.3.5.2 Factors associated with emergency department trajectory groups

The emergency department (ED) model selected satisfied the convergence criterion (GCONV=1E-8) and Likelihood Ratio test (chi-square=178.3998, df=6, p<0.0001). AIC also improved in the model with the intercepts and the predictor variables (AIC=124.574) compared to the intercepts only model (AIC= 284.974). The results of the ordered log odds (logit) regression coefficients are presented in table 12. The estimated log odds of high vs. medium and low ED utilization when all predictor variables were estimated at zero was -6.6242 (p=0.0006) while the estimated log odds of high and medium vs. and low clinic utilization when the predictor variables were estimated at zero was not significant (estimate=-1.3162, p=0.4453).

Race (p=0.0008) had a significant Wald test. However, the 95% CI around the ORs for race approached zero and the odds were too small (OR=0.007; 95% CI, <0.001-

0.128). The explanation for the observed values could be accounted for by the very small numbers in some of the cells corresponding to the different categories of race (Black=328, other=8, unknown=3). As a result, we considered race to be a non-significant predictive of ED group membership.

On the other hand, distance ($p=0.0001$) was predictive of ED group membership. Chronic transfusion (defined as more than 6 annual transfusions) at age 19 ($p<0.001$), diagnosis of chronic pain ($p<0.0001$), being on long-acting narcotics ($p<0.001$) and having AVN of the hip and/or shoulder ($p<0.0001$) were significant sickle cell complications that predicted ED group membership. Having major depression ($p<0.0001$) and anxiety and depression ($p=0.0155$) were significant comorbidities that predicted ED utilization group. Finally, EDR score at age 19 ($p=0.0004$) predicted ED group membership. Table 12 lists the parameters and their corresponding Wald test, while table 13 lists the odds ratios for each of the significant predictors with their corresponding 95% confidence interval.

Individual characteristic.

Disease specific measures. Very high odds ratios and wide 95% confidence intervals were observed for chronic transfusion at 19 (OR =200.376; CI=16.2925->999.999) and chronic pain (OR=211.979; 95%CI, 15.097->999.999). As participants moved from not receiving chronic transfusions to receiving chronic transfusions at age 19, the odds of

high ED utilization were 200.376 times greater than for the combined medium and low ED utilization groups, given that all other predictors were held constant. Similarly, as participants moved from not receiving chronic transfusions to receiving chronic transfusions at age 19, the odds of middle and high vs. low ED utilization were 200.376 times greater, given that all other predictors were held constant.

Being diagnosed with chronic pain also was a significant predictor of ED utilization group membership. When participants had a chronic pain diagnoses, the odds of high ED utilization were 211.979 times greater than for the combined medium and low ED utilization groups, given that all other predictors were held constant. Similarly, as participants moved from no diagnosis of chronic pain to a diagnosis of chronic pain, the odds of middle and high vs. low ED utilization were 211.979 times greater, given that all other predictors were held constant.

AVN of hip and/or shoulder had an odds ratio of 28.866 (95%CI, 5.921-140.731). For participant who had a diagnosis of AVN of the hip and/or shoulder, the odds of high ED utilization were 28.866 times greater than for the combined medium and low ED utilization groups, given that all other predictors were held constant. Similarly, the odds of middle and high vs. low ED utilization were 28.866 times greater when participants had a diagnosis of AVN of the hip and/or shoulder, given that all other predictors were held constant.

Long term narcotics were significant predictors of ED utilization group membership (OR=32.506; 95%CI, 7.717-136.927). For participants on long-acting narcotics, the odds of high ED utilization were 32.506 times greater than for the combined medium and low ED utilization groups, given that all other predictors were held constant. Similarly, the odds of middle and high vs. low ED utilization were 32.506 times greater when participants were on long acting narcotics, given that all other predictors were held constant.

Two comorbidities were significant predictors for ED group membership: diagnosis of major depression (OR=22.54; 95%CI=4.952-102.591) and diagnosis of anxiety and depression (OR=0.126; 95%CI=0.024-0.675). As participants were diagnosed with major depression, the odds of high ED utilization were 22.54 times greater than for the combined medium and low ED utilization groups, given that all other predictors were held constant. Similarly, as participants were diagnosed major depression, the odds of middle and high vs. low ED utilization were 22.54 times greater, given that all other predictors were held constant.

For participants who had a diagnosis of anxiety and depression, the odds of high ED utilization were 0.126 times less than for the combined medium and low ED utilization groups, given that all other predictors were held constant. Similarly, as participants were diagnosed anxiety and depression, the odds of middle and high vs.

low ED utilization was 0.126 times less, given that all other predictors were held constant. Although statistically significant, the clinical implications of such a small odds ratio are questionable.

Contextual factors. While we could not examine the effect of type of health insurance on ED group membership, distance between the participant's location of residence and the sickle cell clinic was a significant predictor of ED group membership (OR=0.948; 95% CI, 0.923-0.974). For every one-mile increase in distance between the home and the clinic, the odds of high ED utilization group were 0.948 times less than for the combined medium and low ED utilization groups, given that all other variables were held constant. Likewise, for every one-mile increase in distance, the odds of middle and low ED utilization groups are 0.948 times lower than for the high ED utilization group.

Transfer related factors. EDR score at 19 had a very large odds ratio and a very wide 95% confidence interval (OR=212.315; 95%CI=10.926->999.999). As EDR increased by one point, the odds of high ED utilization were 212.315 times greater than for the combined medium and low ED utilization groups, given that all other predictors were held constant. Similarly, as EDR increased by one point, the odds of middle and high vs. low ED utilization were 212.315 times greater, given that all other predictors were held constant.

Table 12: Parameter estimates and their corresponding Wald test for hospital model

Analysis of Maximum Likelihood Estimates

Parameter		DF	Estimate	Standard Error	Wald Chi-Square	Pr > ChiSq
Intercept	High ED	1	-6.6242	1.9234	11.8614	0.0006
Intercept	Medium ED	1	-1.3162	1.7242	0.5827	0.4453
AVN of hip and/or shoulder		1	3.3627	0.8083	17.3087	<.0001
Anxiety and depression		1	-2.0684	0.8549	5.8536	0.0155
Major depression		1	3.1153	0.7732	16.2327	<.0001
EDR at age 19		1	5.3581	1.5138	12.5285	0.0004
Long acting narcotics		1	3.4814	0.7337	22.5157	<.0001
Chronic pain		1	5.3565	1.3480	15.7903	<.0001
Chronic transfusion at age 19		1	5.3002	1.2609	17.6684	<.0001
Distance		1	-0.0533	0.0140	14.5237	0.0001
Race		1	-4.9449	1.4738	11.2582	0.0008

Table 13: Odds Ratio Estimates and corresponding 95% confidence interval

Odds Ratio Estimates				
Effect	Point Estimate	95% Wald Confidence Limits		
AVN of hip and/or shoulder	28.866	5.921	140.731	
Anxiety and depression	0.126	0.024	0.675	
Major depression	22.540	4.952	102.591	
EDR at age 19	212.315	10.926	>999.999	
Long acting narcotics	32.506	7.717	136.927	
Chronic pain	211.979	15.097	>999.999	
Chronic transfusion at age 19	200.376	16.925	>999.999	
Distance	0.948	0.923	0.974	
Race	0.007	<0.001	0.128	

4.4 Discussion

This longitudinal database study examined the trajectories of health care utilization for AYAs with SCD during health care transition. We also examined the individual, contextual, and transition related factors that were associated with membership in the different trajectory groups.

Most AYAs who used the sickle cell services at our center had low clinic, hospital, and ED utilization trajectories. Although a small group of participants had high utilization trajectories in the hospital and/ or emergency department, these encounters usually account for the highest cost of care. The sample mean EDR scores were also

below the cutoff point of 0.33 for high ED reliance. However, we did find evidence of increasing reliance on the ED compared to ambulatory care with increasing age indicating that a small group of AYAs with SCD might be at increased risk for high reliance on the ED. Our findings on EDR scores are similar to those of Blinder et al (2015) who studied the age-related EDR score in a large sample of 3208 patients with sickle cell disease and found that beginning at age 15, EDR increased steadily into adulthood. Similarly, Hemker et al. (2011) found that transitioning AYAs had increased EDR scores compared to children and adults. Future research should examine the characteristics of AYAs with high EDR scores to help identify those who might be at risk of increased emergency department reliance and to develop interventions targeted at decreasing their reliance on the ED for care.

Several individual, contextual, and transfer related factors were significant predictors of health care utilization groups in the different services and these factors varied among services indicating the complexity of health care utilization in AYAs with SCD and the factors influencing them. Receiving hydroxyurea was the only common predictor between higher clinic and higher hospital utilization trajectory groups, while distance to the sickle cell center was the only common predictor between higher clinic and higher ED utilization trajectory groups. Depression, chronic pain, long-acting

narcotics, and chronic transfusion at age 19 predicted higher hospital and higher ED trajectory utilization groups.

Our finding with regards to the association between distance from clinic and ED and health care utilization contrasts with other research that found no association between the location of residence and utilization of health care services (Shankar et al., 2005) or found that patients living further from a self-identified provider had higher rates of ED visits (Wolfson, Schragger, Khanna, Coates, & Kipke, 2012). One explanation to our finding could be that the participants in our sample used EDs outside of Duke Health for their emergency care, which were not captured in our database encounters.

Similar to other research we found that chronic pain was a significant predictor health care utilization trajectories. Chronic pain was a prevalent (n=117, 34.51%) and significant predictors associated with both hospital and ED utilization groups, but not clinic groups. Similarly, Sil et al (2016) found that young adults with chronic pain had significantly higher self-report of inpatient hospitalizations compared to those with episodic pain. Studies that examined care seeking behavior for pain in young adults with sickle cell disease found that patients tended to attempt to manage the pain at home and wait until the pain intensity is severe before seeking care (Jenerette, Brewer, & Ataga, 2014). A more aggressive pain management plan and the establishment of outpatient chronic pain management programs might help decrease patient's reliance on

hospital and ED and increase the utilization of outpatient services for the management of acute and chronic pain.

Chronic transfusion at age 19 was also associated with increased hospital and ED utilization in our sample. Similarly, Aisiku et. Al (2009) found that transfusions were associated with high ED utilization. Related to chronic transfusions, iron overload was found to be predictive of increased clinic utilization, but not ED or hospital utilization.

Our finding that gender was not a significant predictor of any of the health care utilization groups is similar to the study by McClish et al. (2006). Our finding that as participant's age increased the odds of high clinic utilization were greater compared to medium and low clinic utilization and the odds of high clinic and medium clinic utilization were greater compared to low clinic utilization is similar to other research. A study that compared older adults with SCD to younger adults with SCD on ED and clinic utilization found that older adults tended to use the clinic more for pain management and other SCD complications (Sanders, Labott, Molokie, Shelby, & Desimone, 2010). However, unlike other research which found that patients who were 21 years of age or older had higher ED visits than younger patients (Wolfson, Schragar, Khanna, Coates, & Kipke, 2012), age was not a significant predictor for neither ED nor hospital utilization group membership in this sample. One explanation could be that

AYAs utilized ED and hospitals outside of our network for their care and our database could not capture these encounters.

It was surprising that SCD genotype, severity score at ages 15 and 19, and the total number of VOC and ACS at ages 12 to 27 were not predictive of group membership in any of the health care utilization categories. This is despite the prevalence of VOC (n=271, 79.94%) and ACS (n=138, 40.71%) in the sample. Our findings are different from other studies which found that SCD severity was associated with high ED encounters (Aisiku et al., 2009) and return visits to the ED (Akenroye et al., 2014), but similar to Epstein et al. (2006) who found no association between SCD genotype and emergency department encounters in adult patients with SCD. One explanation to our findings could be that the modification we made to the sickle cell severity index affected our result to accurately measure severity. We intent to conduct a factor analysis of the modified severity index we used in the study to determine its validity in the modified form.

A major finding in our study was the association between health care utilization and mental health conditions. Depression was a prevalent (n=63, 18.58 %) and significant predictor of utilization group membership for the hospital and ED. The odds of being in the high utilization groups for hospital and ED were greater compared to the medium and low utilization groups if depression was present. Sandoval et al. (2010)

interviewed two groups of patients: one with frequent visits to the ED and those without and found that those who had frequent ED visits reported higher levels of stress and were more likely to screen positive for depression (Sandoval et al., 2010). Moreover, four participants in this study attempted suicide, one of whom attempted suicide four times. There was no obvious pattern in terms of age, but it appeared that some of the characteristics these participants shared were SS genotype, high disease severity, high ED utilization, medium to high hospital and clinic utilization, diagnosis of depression, diagnosis of chronic pain, and diagnosis of cerebral infarct. Although four participants are not a large number and could be an under estimation of the actual suicidal ideation in the sample since we relied on ICD 9 codes for a diagnosis of suicidal ideation, the implications from this finding are significant given the prevalence of depression in this sample and the high prevalence of depression in sickle cell disease in general (Benton, Ifeagwu, & Smith-Whitley, 2007; Jerrell, Tripathi, & McIntyre, 2011). These findings have major implications for clinical practice and emphasize the need to frequently screen AYAs with severe form of SCD, chronic pain, cerebral infarcts, and high utilization for depression and suicidal ideation.

One goal of our study was to describe successful transition. Transfer to adult care has been considered as a measure of successful transition, yet more comprehensive measures of transition success are needed to identify AYAs who might be struggling

with the transition process. Successful transition process ensures uninterrupted access to primary and subspecialist care for the AYAs with SCD, promotes independent self-care skills, nurtures social and emotional development through teaching advocacy and communication skills, and provides a sense of support for future health and life goals (McPheeters et al., 2014). The majority of our sample has transferred to adult care. The mean age at transfer was 19 years. We also examined the continuity of care after transfer and the majority of participants who transferred to adult care had at least one additional encounter in the adult sickle cell clinic, and around 65% had more than ten encounters. Only a few (5.83%) had no adult encounters after transfer. As a result, we consider the vast majority of participants who had at least one encounter in the adult sickle cell clinic to have successfully transferred and integrated into adult care.

4.5 Limitations

As with any secondary data analysis, we were limited to the variables available in the dataset. However, we were able to obtain or create almost all variables of interest from data already available in the database with the exception of participant health insurance and participant's laboratory values due to missing data. Although several studies have found an association between type of insurance and health care utilization (Brousseau, Owens, Mosso, Panepinto, & Steiner, 2010; Mvundura, Amendah, Kavanagh, Sprinz, & Grosse, 2009; Schlenz, Boan, Lackland, Adams, & Kanter, 2016) we

were unable to make any association between health care utilization and the type of health insurance because of missing data.

A second limitation is that our analyses of encounters were limited to encounters within Duke Health and did not account for health care utilization that occurred at other health care institutions. Third, ICD 9 accuracy could be an issue in secondary database analyses. We attempted to verify and address any ICD9 code inaccuracy in the dataset during data cleaning by examining the ICD9 code and the corresponding diagnoses name for congruence.

4.6 Conclusion and Implications

To our knowledge, this is the first longitudinal study that examined the trajectories of health care utilization in transitioning AYAs with SCD and the conducted a comprehensive exploration of factors associated with these trajectories. Individual, contextual, and transfer related factors were associated with the different health care utilization trajectory groups for AYAs with SCD. A major finding in this study was the association of several mental health conditions, including depression, with health care utilization group membership. This finding underscores the importance of routine screening and adequate management of mental health care conditions in AYAs with SCD, especially during transfer to adult care.

4.7 Acknowledgement

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5. Health Care Transition in Adolescents and Young Adults with Sickle Cell Disease: Future Directions

The purpose of this dissertation was to provide a better understanding of health care transitions in adolescents and young adults (AYAs) with sickle cell disease (SCD). This purpose was achieved through an examination of the state of the science on health care transition in AYAs with SCD, the challenges of shifting self-management from the parent to the adolescents with SCD prior to transition to adult care, and the patterns of health care utilization during transition and the factors associated with these patterns.

5.1 Summary of Findings

Our integrative review of original research on health care transition in AYAs with SCD revealed major gaps in the literature with regards having an adequate understanding of how AYAs take on more disease management responsibility, and how does health care transition impact AYAs outcomes. Overall, very few studies examined health care transition outcomes in terms of acquiring independence and integration into adult care, what is considered successful transition, and factors that would facilitate or hinder successful transition. Finding from the review also indicated that assuming more care responsibility by the AYA with SCD was a critical and challenging process.

Our qualitative descriptive study examining the shifting of management responsibility from the parent to the adolescent with SCD aims to address the current gap in the literature. For the adolescents in our study, complex unpredictable symptoms

made it difficult to take on the self-management responsibility. The addition of the non-normative stressors of complex SCD management layered on top of normative developmental challenges and transitions faced by adolescents (Kennedy, Gask, & Rogers, 2005). The adolescents also struggled with maintaining control over the events in their lives because of the unpredictability of SCD. This challenge is particularly significant given that achieving autonomy and independence are important developmental milestones for adolescents. The adolescents addressed this challenge with adaptive work directed at having more autonomy over their management, including understanding how SCD affected them, pushing back at parents and over time stepping up for more decision making responsibility.

The adolescents' struggle with managing complex symptoms was mirrored by parents' hesitation to fully shift the management responsibility to the adolescent, because parents felt that the adolescent lacked their level of expertise in SCD management. Mentoring the adolescent to master disease management and triangulate different knowledge sources as the parents had done over the years constituted a challenge for the parents, particularly amidst an unpredictable disease trajectory. The parents also worried that the adolescent might be willing to take "extreme" risks for better quality of life. This situation made it difficult for them to shift the management decision making to the adolescent because they felt the need to remain in control to

ensure the adolescent's wellbeing. Perhaps the greatest challenge for the parents was to balance fostering the adolescents' independence with maintaining their wellbeing. In the case of SCD this challenge comes with a grave cost: hospitalization and sickle cell crises. The parents' adaptive work to address these challenges included engaging the adolescent in discussions about their condition and how they are responding to it as well as collaborating with the adolescent on SCD management.

However, collaboration and goal-setting across the adolescents- parent-provider triad will assist in facilitating the progressive shift of self-management responsibility from parents to adolescents and thus improve outcomes for adolescents with SCD and their parents.

The purpose of our longitudinal study was to address another gap in the literature and was to identify the different trajectories of health care utilization of transitioning AYAs with SCD and the factors associated with membership in these trajectory groups. Our findings from the longitudinal examination of health care utilization for AYAs with SCD during transition to adult care revealed clear utilization typology across the sickle cell clinic, hospital, and emergency department (ED). The majority of patients who used sickle cell services had low clinic, hospital, and ED utilization trajectories. A little over 20% of participants had medium utilization trajectories in at least one of the services and around 6-11% had high utilization in at

least one of the services. Low clinic utilizers tended to have low ED and hospital utilization trajectories. A small group of medium & high clinic utilizers had both high hospital and high ED utilization trajectories.

We also examined transfer and integration into adult care for our sample. The mean age at transfer in our sample was 19 years and the vast majority of participants who transferred to adult care had at least one additional encounter in the adult sickle cell clinic. Around 65% had more than ten encounters and only a few patients (5.83%) had no adult encounters after transfer. As a result, we consider the vast majority of participants who had at least one encounter in the adult sickle cell clinic to have successfully transferred and integrated into adult care.

We then examined the factors associated with group membership in each of the service trajectories. Significant predictors for clinic group membership included individual and contextual factors. Several sickle cell complications were also significant, including cerebral infarction, iron overload, the number of blood transfusions at age 19, lower extremity ulcer, and being on hydroxyurea. Attention deficit disorders were the only significant comorbidity that predicted of sickle cell clinic utilization trajectories. Finally, age at the time of transfer to adult care was a significant transfer related factor that predicted clinic group membership.

No significant contextual, or transfer related factors predicted hospital group membership, while several sickle cell complications were significant predictors, namely chronic transfusion at age 19, diagnosis of chronic pain, and being on hydroxyurea and long-acting narcotics. Being diagnosed with depression and having intellectual disabilities were significant comorbidities that predicted hospital utilization trajectory group as well.

Finally, distance was a predictor contextual factor for ED group membership. Chronic transfusion at age 19, diagnosis of chronic pain being on long-acting narcotics, and having AVN of the hip and/or shoulder were significant sickle cell complications that predicted ED group membership. Having major depression and anxiety and depression were significant comorbidities that predicted ED utilization group membership. EDR score at age 19 was a predictor transfer related variable that predicted ED utilization group membership as well.

It was surprising that SCD genotype, severity score at ages 15 and 19, and the total number of VOC and ACS at ages 12 to 27 were not predictive of group membership in any of the health care utilization categories. This is despite the prevalence of VOC (n=271, 79.94%) and ACS (n=138, 40.71%) in the sample. Our findings are different from other studies which found that SCD severity was associated with high ED encounters (Aisiku et al., 2009) and return visits to the ED (Akenroye et al., 2014), but similar to

Epstein et al. (2006) who found no association between SCD genotype and emergency department encounters in adult patients with SCD. One explanation to our findings could be that the modification we made to the sickle cell severity index affected our result to accurately measure severity. We intent to conduct a factor analysis of the modified severity index we used in the study to determine its validity in the modified form. Nonetheless, several SCD complications were predictive of health care utilization patterns in our study. Chronic pain was a prevalent (n=117, 34.51%) and significant predictors associated with both hospitals and ED utilization groups, but not clinic groups.

Chronic transfusion at age 19 was also associated with increased hospital and ED utilization in our sample. Similarly, Aisiku et. Al (2009) found that transfusions were associated with high ED utilization. Related to chronic transfusions, iron overload was found to be predictive of increased clinic utilization, but not ED or hospital utilization.

A major finding in our study was the association between health care utilization and mental health conditions. Depression was a prevalent (n=63, 18.58 %) and significant predictor of utilization group membership for the hospital and ED. The odds of being in the high utilization groups for hospital and ED were greater compared to the medium and low utilization groups if depression was present. Sandoval et al. (2010) interviewed two groups of patients: one with frequent visits to the ED and those without

and found that those who had frequent ED visits reported higher levels of stress and were more likely to screen positive for depression (Sandoval et al., 2010).

Moreover, four participants in this study attempted suicide, one of whom attempted suicide four times. There was no obvious pattern in terms of age, but it appeared that some of the characteristics these participants shared were SS genotype, high disease severity, high ED utilization, medium to high hospital and clinic utilization, diagnosis of depression, diagnosis of chronic pain, and diagnosis of cerebral infarct. Although four participants are not a large number, the implications from this finding are significant given the prevalence of depression in this sample and the high prevalence of depression in sickle cell disease in general (Benton, Ifeagwu, & Smith-Whitley, 2007; Jerrell, Tripathi, & McIntyre, 2011). Patients with severe form of sickle cell disease, chronic pain, and cerebral infarcts, and high utilization should be frequently screened for depression and suicidal ideation.

5.2 Limitations

Recruitment and sampling for our focus groups was limited to one health system, which might limit the external validity of the study. However, Duke Health is a major referral center in the southeast United States, and participants from a geographical radius of up to 100 miles were in our sample. Our sample was mostly African Americans. However, this is expected given that SCD occurs predominately in African Americans in

the United States (Hassell, 2010). Our adolescent sample consisted mostly of male participants, with only three female adolescents. Yet, our adolescent sample was similar to reported population estimates in the mean number of hospital admissions and emergency department use (Paulukonis et al., 2014). In addition, our parent sample had at least a high school education and consisted mostly of female caregivers. The educational level of our parent sample might limit the generalizability of our findings to other parent samples. The fact that the majority of the adolescents' primary caregivers were female was not surprising; studies report on the important role that female caregivers play in the care of the chronically ill in the African American population (Becker, Gates, & Newsom, 2004; Jenerette & Valrie, 2010). Despite these limitations, this study contributes in-depth understanding of the challenges that adolescents with SCD and their parents face in shifting the management responsibility from the perspectives of adolescents and parents.

As with any secondary data analysis, our longitudinal study was limited to the variables available in the dataset. However, we were able to obtain or create almost all variables of interest from data already available in the database with the exception of participant health insurance and participant's laboratory values due to missing data. Although several studies have found an association between type of insurance and health care utilization (Brousseau, Owens, Mosso, Panepinto, & Steiner, 2010;

Mvundura, Amendah, Kavanagh, Sprinz, & Grosse, 2009; Schlenz, Boan, Lackland, Adams, & Kanter, 2016) we were unable to make any association between health care utilization and the type of health insurance because of missing data.

A second limitation is that our analyses of encounters were limited to encounters within Duke Health and did not account for health care utilization that occurred at other health care institutions. Third, ICD 9 accuracy could be an issue in secondary database analyses. We attempted to verify and address any ICD9 code inaccuracy in the dataset during data cleaning by examining the ICD9 code and the corresponding diagnoses name for congruence.

5.3 Future direction for research and practice

Vygotsky emphasized the role of social forces, including parental guidance and interaction, teacher instruction, and language in child development (Miller, 1993). Within this framework, enhancing self-management capacity in the adolescent requires an iterative process of parent-adolescent communication. It might be easier for everyone concerned to maintain the status quo with the parents in full charge of the management, particularly if their strategies have been successful in maintaining the adolescent relatively healthy. While this approach might be effective when the adolescent is residing with the parent, self-management becomes a challenge when the adolescent leaves the home, for example, to college. Health care providers need to collaborate with

parents and adolescents to incrementally shift the responsibility of disease management (Young et al., 2014). Health care providers need to assess the parent-child relationship and their progress in shifting the management responsibility, facilitate discussions to arrive on a shared understanding of the challenges they are having with shifting the management responsibility, and collaborate on adaptive work to address these challenges.

Our study described the challenges that adolescent with SCD and their parents face with shifting the management responsibility. Further research should explore the challenges adolescents with other chronic conditions and their parents face in shifting the management responsibility to understand the challenges across conditions and the unique challenges specific to certain conditions, including those involving neurological sequelae or cognitive deficits. Further research should also assess the adolescent's readiness and their competency in taking on more management responsibility as well as develop and test interventions aimed at facilitating the shifting of management responsibility from the parent to the adolescent with chronic illness.

To our knowledge, our longitudinal study was the first study that examined the trajectories of health care utilization in transitioning AYAs with SCD and the conducted a comprehensive exploration of factors associated with these trajectories. Individual, contextual, disease, comorbidities, and transfer related factors were associated with the

different health care utilization trajectory groups for AYAs with SCD. A major finding in this study was the association of several mental health conditions, including depression, with health care utilization group membership. This finding underscores the importance of routine screening and adequate management of mental health care conditions in AYAs with SCD, especially during transfer to adult care.

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Biography

Mariam Kayle was born in Beirut, Lebanon on September 6th, 1978. She graduated with Bachelors of Science in Nursing, with distinction, from the American University of Beirut (1999) and Master of Science in Nursing from Duke University (2005). Mariam has coauthored a book chapter titled : “Communicating with acutely ill Children: An individualized approach” and coauthored the following articles in peer-reviewed journals: 1)Challenges in shifting management responsibility from parents to adolescents with sickle cell disease, 2)Evaluation of a Sickle Cell Disease Educational Website for Emergency Providers, 3)The adolescent and young adult with cancer: A developmental life course perspective, 4)Attitudes toward patients with sickle cell disease in a multicenter sample of emergency department, and 5)Survival at the threshold of viability: A nationwide survey of the opinions and attitudes of physicians in a developing country. Mariam has received scholarships from the American University of Beirut Medical Center for her master studies and from the Graduate School at Duke University for her doctoral studies. She also received the Summer Fellowship from the graduate school at Duke University for three consecutive years. Mariam was inducted to Sigma Theta Tau International honor society of nursing in 2005. She is the recipient of many honors including the “Aallamouna" (Ambassador’s)

Profiling Award (Order of Nurses in Lebanon, 2012) and the Mary Crawford Memorial Award (American University of Beirut School of Nursing, 1999).