DECISION-MAKING INVOLVEMENT, SELF-EFFICACY,
AND THE RELATIONSHIP TO TRANSITION READINESS
IN YOUTH WITH SICKLE CELL DISEASE

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By
MAUREEN M. VARTY
Dr. Lori Popejoy, Dissertation Supervisor

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The undersigned, appointed by the dean of the Graduate School, have examined the dissertation entitled:

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RELATIONSHIP TO TRANSITION READINESS IN YOUTH WITH SICKLE CELL
DISEASE

Presented by Maureen M. Varty, a candidate for the degree of doctor of philosophy, and hereby certify that, in their opinion, it is worthy of acceptance.

Dr. Lori Popejoy
Dr. Katherine Patterson Kelly
Dr. Bonnie J. Wakefield
Dr. Russell Ravert
Dr. Barbara Speller-Brown

DEDICATION

This dissertation is dedicated to my husband, Ted Varty. In his daily life, he inspires me to seek knowledge and to stay curious as if it was as important as seeking air. For the past few years he has pursued a full-time career while also seeking to finish a graduate degree of his own. In this, he showed me how to endure sacrifices with grace. Throughout this program, he encouraged me to look at the big picture and seek opportunities which would help me to find happiness in my career. Ted frequently reminded me of his faith in my abilities to be successful in this doctoral program. Often his greatest support came through just listening to me talk about my challenges until I found the answers within my own thoughts. In addition, he provided never-ending assistance as a technical editor. I will be forever grateful for his patience and the endless support of my passions.

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TABLE OF CONTENTS

ACKNOWLEDGEMENTS	ii
LIST OF TABLES	vi
LIST OF FIGURES	vii
LIST OF SUPPLEMENTS	viii
Abstract.....	ix
CHAPTER ONE : GENERAL INTRODUCTION	1
Conclusion.....	4
References	6
CHAPTER TWO: A SYSTEMATIC REVIEW OF TRANSITION READINESS IN YOUTH WITH CHRONIC DISEASE.....	8
Abstract	8
Method	11
Results	14
Discussion	22
References	28
CHAPTER THREE: YOUTHS' EXPERIENCES OF TRANSITION FROM PEDIATRIC TO ADULT CARE: AN UPDATED QUALITATIVE METASYNTHESIS	43
Abstract	43
Methods.....	47
Results	50
Discussion	64
Conclusions	67

References	68
CHAPTER FOUR: METHOD	99
Specific Aims	99
Specific Aim 1:	101
Specific Aim 2:	102
Specific Aim 3:	102
Research Strategy	102
Approach	113
References	124
CHAPTER FIVE: DECISION-MAKING INVOLVEMENT, SELF-EFFICACY,	
AND THE RELATIONSHIP TO TRANSITION READINESS IN YOUTH WITH	
SICKLE CELL DISEASE	130
Abstract	130
Methods	135
Results	138
Discussion	141
Conclusion	146
References	148
CHAPTER 6: CONCLUSION.....	159
Dissertation Overview and Findings	160
Discussion	163
Future Research	165
References	169
COMPREHENSIVE REFERENCE LIST.....	172
VITA.....	192

LIST OF TABLES

Table 1. Quality Assessment of Methodology for Included Studies	39
Table 2. Demographic and Ecology Factors Found to have Significant or Inconsistent Relationships to Transition Readiness	40
Table 3. Psychosocial Factors Found to have Significant or Inconsistent Relationships to Transition Readiness	41
Table 4. Self-Management and Transition Education Factors Found to have Significant or Inconsistent Relationships to Transition Readiness	42
Table 5. Main Themes and Definitions.....	76
Table 6. Deficiencies and Recommendations Reported by AYA.....	77
Table 7. Description of Variables and Methods for Data Collection	117
Table 8. Description of Analysis Methods to Address Study Aims	121
Table 9. Description of Variables and Measures	154
Table 10. Sample Characteristics.....	155
Table 11. Correlations Between AYA Decision-Making Involvement and Self-Efficacy	156
Table 12. Significant Predictors of AYA Perceived Transition Readiness	156
Table 13. Significant Predictors of AYA Perceived Healthcare Responsibility	157
Table 14. Significant Predictors of Family Caregiver Perceived AYA Transition Readiness	157
Table 15. Significant Predictors of Family Caregiver Perceived AYA Healthcare Responsibility	158

LIST OF FIGURES

Figure 1. PRISMA Flow Diagram for the Systematic Review.....	38
Figure 2. PRISMA Flow Diagram for the Metasynthesis.....	75

LIST OF SUPPLEMENTS

Supplement 1. Characteristics Across Studies.....	78
Supplement 2. CASP Appraisal of Included Studies.....	90
Supplement 3. CERQual Qualitative Evidence Table	92

Decision-Making Involvement, Self-Efficacy, and the Relationship to Transition
Readiness in Youth with Sickle Cell Disease

Maureen M. Varty, BSN, RN

Dr. Lori L. Popejoy, Dissertation Advisor

Abstract

Transition from pediatric to adult healthcare settings poses significant risks to morbidity and mortality for adolescents and young adults with sickle cell disease. Increasing transition readiness, the acquisition of disease-specific knowledge and self-management skills, has been proposed as strategy to improve health outcomes for adolescents and young adults (AYA) after transition. To date, few studies have looked at factors that may influence transition readiness, a measure of disease-specific knowledge and self-management, in AYAs with sickle cell disease. This limits healthcare providers' and researchers' abilities to develop interventions specific to the unique needs of the population. This dissertation study examined the relationships between decision-making involvement, self-efficacy of sickle cell disease self-management, and transition readiness in AYAs with sickle cell disease prior to transition to adult healthcare settings. This study found that higher levels of expressive behaviors such as sharing opinions and ideas in decision-making were associated with higher levels of AYA healthcare responsibility. Self-efficacy was positively associated with transition readiness but inversely related to AYA healthcare responsibility. Parent involvement was negatively associated with AYA healthcare responsibility. Future research interventions that increase AYA involvement in decision-making regarding disease management, increase

self-efficacy, and safely reduce parent involvement may positively influence their capacity for self-management.

CHAPTER ONE

GENERAL INTRODUCTION

Sickle cell disease (SCD) is an autosomal recessive disorder which affects hemoglobin, an oxygen-carrying protein in red blood cells (Brousse, Makani, & Rees, 2014; National Heart, Lung, and Blood Institute [NHLBI], 2018). Those living with SCD produce hemoglobin *S*, an abnormal hemoglobin, which tends to make red blood cells sticky and crescent-shaped. (NHLBI, 2018). In the United States, a majority of affected individuals are African-American; however, worldwide, SCD impacts individuals of Indian, Middle Eastern, Mediterranean, Caribbean, South and Central Americans, and Chinese ethnicity (NHLBI, 2018).

Due to differences between individuals and the varieties of SCD, symptoms and complications differ in severity. Symptoms and complications of SCD are primarily caused by hemolysis and vaso-occlusions. Hemolysis is reflected in lower hemoglobin levels with the level of anemia varying across the different SCD types (NHLBI, 2018). Lower hemoglobin levels lead to a variety of symptoms including fatigue, jaundice, slowed growth, and delayed puberty (Centers for Disease Control and Prevention, 2019). Vaso-occlusions occur because the sickle-shaped red blood cells tend to stick to each other and blood vessel walls (NHLBI, 2018). For individuals living with SCD, these vaso-occlusions are most commonly experienced as acute pain crises ranging from mild to severe (Ballas, 2011; NHLBI, 2018). A majority of acute pain crises are managed at home (Telfer & Kaya, 2017); however, when self-care strategies at home are ineffective then individuals are encouraged to seek medical care (Jenerette, Brewer, & Ataga, 2014). The onset of pain is associated with dehydration, stress, weather changes, cold exposure,

overexertion, and fatigue (Ballas, 2011). As youth age they often begin to experience chronic pain, and this pain can be exacerbated by repeated acute pain crises (NHLBI, 2018). Chronic pain is experienced between acute pain crises; however, it is reported as milder in nature (Ballas, 2011). Though the etiology of the chronic pain is not completely understood, it thought to be caused by inflammatory, neuropathic, and ischemic mechanisms (Brandow, Zappia, & Stucky, 2017). There can be objective sources for this pain including avascular necrosis of the femoral and humoral heads, leg ulcers, and bony infarcts; however, there is not always an objective source (Brandow et al., 2017). In addition to acute pain crises, SCD vaso-occlusion causes damage to the brain, spleen, liver, lungs, kidneys, heart, bones, skin, penis, and joints over a person's lifetime (NHLBI, 2018). Severe, life-threatening complications of SCD include splenic sequestration crises, aplastic crises, infections, acute chest syndrome, and strokes (NHLBI, 2018). Treatment for SCD is focused on preventing and addressing complications associated with SCD as they develop over time (NHLBI, 2018).

The Necessity and Risk of Transition to Adult Healthcare

As of 2006, a blood test for SCD is included in routine newborn screening; this has increased early detection of SCD and has helped to prevent complications (Benson & Therrell, 2010). Advances in medical treatment, coupled with improved screening for SCD, have dramatically increased the life expectancy of those living with SCD from 14 years in 1973 to 40-60 years today (Elmariah et al., 2014). With over 93% of youth living into adulthood, a majority of young adults living with SCD need to transition to the adult healthcare system to ensure continuity of care (Quinn, Rogers, McCavit, & Buchanan, 2010). Transition is a critical event as there are increased risks for complications for

young adults living with SCD after transition. Immediately post-transition, young adults with SCD experience increased rates of rate of acute care visits due to SCD complications (Dickerson, Klima, Rhodes, & O'Brien, 2012; Quinn et al., 2010). There is also an inappropriate reduction in the use of routine medical treatment which further increases their risk of complications such as acute pain crises, strokes, and acute chest syndrome (Blinder et al., 2013). Most concerning is the increase in mortality just after transition (Blinder et al., 2013). Improving transition processes and transition readiness has received increased attention in recent years because successful transition may reduce adverse outcomes and minimize disruptions in treatment.

Current Research on Transition Readiness

Transition readiness is the acquisition of disease-specific knowledge and self-management skills required by adolescents and young adults (AYA) to successfully transition from pediatric to adult healthcare. To date, few studies have identified factors that may influence transition readiness in AYAs with sickle cell disease (Varty & Popejoy, 2020). Two non-modifiable factors, age and SCD severity, are found to be significantly related to transition readiness, specifically as an AYA ages, transition readiness increases ($p = 0.003$) and more severe SCD resulted in less interest in activities related to transition readiness ($p = 0.043$) (McPherson, Thaniel, & Minniti, 2009). The only modifiable factor which has been found to be significantly related to transition readiness in AYAs living with SCD is self-efficacy (Treadwell et al., 2016). Self-efficacy in SCD self-management is the confidence the AYA has in their ability to manage their disease (Edwards, Telfair, Cecil, & Lenoci, 2000). As self-efficacy increased so does transition readiness (Treadwell et al., 2016). Self-efficacy is positively related to

transition readiness and is used as outcome of transition preparation. As AYAs with SCD develop knowledge and skills for disease self-management, it is expected that not only is their transition readiness improving but so is their confidence in their ability to care for their disease. As self-efficacy is the only modifiable factor known to be associated with transition readiness for AYAs with SCD, further research is necessary to identify factors related to transition readiness and to guide intervention development.

Conclusions

The purpose of this dissertation project was to assess AYA decision-making involvement, a novel factor in the area of transition readiness, for its relationship to self-efficacy and transition readiness in AYAs with SCD. AYA decision-making involvement is the way in which AYAs contribute to the decision-making process with their parents regarding the daily management of their disease (Miller & Drotar, 2006). Increasing AYA decision-making involvement has been found to be positively related to self-efficacy and disease self-management behaviors such as treatment adherence (Liprie, 1993; Miller & Drotar, 2006; White, 1996). This relationship has not been studied in AYAs living with SCD. This research assessed this gap in knowledge regarding the relationships between decision-making involvement, self-efficacy, and transition readiness in AYAs living with SCD. It was hypothesized in this study that decision-making involvement would be positively related to self-efficacy and transition readiness. It was also hypothesized that, as the AYA engaged more in decision-making regarding their disease, they would have higher confidence in their knowledge and skills related to disease self-management. Identifying modifiable factors associated with transition

readiness is crucial to developing better interventions. Findings from this study will be used to inform future intervention development and research.

The subsequent chapters in this dissertation (Chapters 2-6) are all components of this dissertation project. Chapter two is a systematic review of factors associated with transition readiness in AYAs living with chronic disease. This review encompassed a variety of chronic diseases and was used to inform the design of this dissertation project by identifying which factors had been researched in AYAs with SCD and which factors still merited further study. Chapter three is a metasynthesis which explored the experiences and expectations of AYAs living with a chronic disease on transition preparation and transition itself. This metasynthesis encompassed a variety of chronic diseases, and the literature included was from 2011-2019. The metasynthesis complemented the review of literature and identified that AYAs believed parents had a role in transition preparation and the development of transition readiness. Finding from the review and metasynthesis informed the research design and methods for the dissertation project. Chapter four is the research proposal supporting this work and was funded by the National Institute of Nursing Research Ruth L Kirchstein Predoctoral Individual National Research Service Award for 2019-2020. Chapter five is the dissertation study procedures and findings. Finally, Chapter six provides a synthesis of the overall dissertation project, the findings, and the significance to the profession of nursing.

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CHAPTER TWO

A SYSTEMATIC REVIEW OF TRANSITION READINESS IN YOUTH WITH CHRONIC DISEASE

Varty, M., & Popejoy, L. L. (2020). A Systematic Review of Transition Readiness in Youth with Chronic Disease. *42*(7), 554-566. doi: 10.1177/0193945919875470

Abstract

The transition of chronically ill adolescents and young adults to adult health care is poorly managed, leading to poor outcomes due to insufficient disease knowledge and a lack of requisite skills to self-manage their chronic disease. This review analyzed 33 articles published between 2009 and 2019 to identify factors associated with transition readiness in adolescents and young adults with chronic diseases, which can be used to design effective interventions. Studies were predominantly cross-sectional survey designs that were guided by interdisciplinary research teams, assessed adolescents and young adults ages 12–26 years, and conducted in the outpatient setting. Modifiable factors, including psychosocial and self-management/transition education factors, and non-modifiable factors, including demographic/ecological and disease factors, associated with transition readiness were identified. Further research is necessary to address gaps identified in this review prior to intervention development, and there is a need for additional longitudinal studies designed to provide perspective on how transition readiness changes over time.

Keywords: transition readiness, transitions, adolescents, young adults, chronic disease, systematic review

The majority of youth with chronic diseases live into adulthood and must transition to the adult health care setting for continued disease management and health promotion. Transition has been 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,” which implies that strategic steps must be taken to prepare adolescents and young adults for the change in expectations between the pediatric and adult health care setting (Blum et al., 1993). Despite frequent calls to improve transitional care, transition preparation that encourages adolescents and young adults to assume independent responsibility for disease self-management and to make timely changes in their health care team is still inadequate (McManus et al., 2013). Although advancements have been made to improve quality of life and life expectancy for many pediatric chronic diseases, transition to adult health care poses significant risk to continuity of care, disease management, and to the adolescent and young adult’s quality of life and livelihood (McManus et al., 2013; Stam, Hartman, Deurloo, Groothoff, & Grootenhuis, 2006).

Negative outcomes related to poor transition have been reported in many different chronic disease populations including diabetes (Garvey et al., 2012), liver transplantation (Annunziato et al., 2007), sickle cell disease (Blinder et al., 2013; Brousseau, Owens, Mosso, Panepinto, & Steiner, 2010), inflammatory bowel disease (de Silva & Fishman, 2014), chronic kidney disease (Fenton, Ferris, Ko, Javalkar, & Hooper, 2015), human immunodeficiency virus (Fish et al., 2014), congenital heart disease (Goossens et al., 2011), attention-deficit hyperactivity disorder (Montano & Young, 2012), and hydrocephalus (Simon et al., 2009). Studies looking at adherence found that after poor

transitions, there were increased rates of non-adherence in liver transplantation recipients (Annunziato et al., 2007) and increased hemoglobin A1c levels in young adults with type 1 diabetes (Garvey et al., 2012; Helgeson et al., 2013). After transition to adult health care, there is a decrease in routine outpatient care health care utilization (Blinder et al., 2013) and an increase in hospital admissions and emergency room visits (Brousseau et al., 2010; Dickerson, Klima, Rhodes, & O'Brien, 2012). Latent increases in mortality after the transition to adult health care for chronic diseases such as sickle cell disease and human immunodeficiency virus have also been reported (Fish et al., 2014; Quinn, Rogers, McCavit, & Buchanan, 2010). Studies reporting adolescents and young adults' experiences of the transition to adult health care found that they believed they had insufficient knowledge and self-management skills to successfully manage their disease (Asp, Bratt, & Bramhagen, 2015; Burström, Öjmyr-Joelsson, Bratt, Lundell, & Nisell, 2016). In addition, they reported being unable to identify appropriate adult health care providers (Bemrich-Stolz, Halanych, Howard, Hilliard, & Lebensburger, 2015) and found it challenging to establish a trusting patient-provider relationship in their new adult health care setting (Bemrich-Stolz et al., 2015; Garvey et al., 2014; Huang et al., 2011). The issue of trust is crucial as adolescents and young adults are more likely to disengage from health care if they fail to establish a trusted relationship with their adult health care provider similar to what they had experienced in pediatrics (Bemrich-Stolz et al., 2015; Garvey et al., 2014; Lewis & Noyes, 2013). Preparation for transition to adult care should become a priority within pediatric clinical practice in order to positively impact health and utilization outcomes.

Pediatric health care providers must make transition preparation a routine practice by developing and implementing evidence-based approaches to increase the level of adolescent and young adult's disease-specific knowledge and help them develop self-management skills needed for successful transition to adult care (American Academy of Pediatrics, American Academy of Family Practice, & American College of Physicians, 2002). This level of preparation is known as transition readiness. Routine assessment of transition readiness provides a baseline of the adolescent and young adult's current understanding about their condition and their capacity to perform disease self-management (Zhang, Ho, & Kennedy, 2014). Understanding factors associated with transition readiness is an essential first step to developing interventions to positively influence adolescents and young adult's transition readiness. Yet little is known about factors that positively and/or negatively influence transition readiness in adolescents and young adults. To the best of our knowledge, no systematic reviews had been conducted on this topic. The primary purpose of this systematic review was to explore the literature regarding factors associated with transition readiness in adolescents and young adults with chronic diseases and to identify factors that could be used to design effective interventions.

Methods

This systematic review was conducted and reported according to the Preferred Reporting Items for Systematic Reviews and Meta-Analysis (PRISMA) guidelines. A meta-analysis was not completed due to the heterogeneity of the available literature.

Inclusion Criteria

For the purpose of this review, transition readiness was defined as the following: the attainment of disease-specific knowledge and the development of effective self-management skills needed for successful transition from pediatric to the adult health care settings. To be included, a study needed to be a primary, peer-reviewed research that assessed transition readiness in adolescents and young adults aged 12–30 years living with a chronic disease, be available in full-text, and be written in English. Studies were excluded if the chronic disease limited the adolescent's or young adult's ability to become fully independent in disease self-management or were not peer reviewed.

Search Strategy

Databases were searched for years 1990–2019 and included PubMed, Google Scholar, CINAHL, Medline, and PsycINFO. The year 1990 was selected, because it is the first year in which transition was mentioned as a significant issue in the care of adolescents and young adult's health care (Blum et al., 1993). Search strategies and key terms were guided by a medical librarian, and they included restricting the language to English and studies involving humans. Chronic diseases included in the search terms were those common to childhood that continue to require lifelong medical management. Key terms included “transition,” “transition readiness,” “chronic disease,” “chronic illness,” “sickle cell,” “diabetes,” “epilepsy,” “asthma,” “spina bifida,” “cystic fibrosis,” “adolescent,” “teen,” “youth,” and “young adult.”

Article Selection

The search strategy was first trialed in PubMed and PsycINFO; once it was found that the strategy was effective in identifying articles appropriate for inclusion, Google Scholar, CINAHL, and Medline were searched. After identifying potential articles from

PubMed, Google Scholar, CINAHL, Medline, and PsycINFO databases, articles were excluded first by assessing titles and abstracts for duplicates and then applying the inclusion criteria. Next, the remaining full-text articles were read to ensure that inclusion criteria were met, and if so, then the articles were assessed for quality. Bibliographies of the articles were also screened for potential articles that may meet the inclusion criteria. These articles were then located and screened for inclusion and quality appraisal. The article selection process is detailed in Figure 1.

Quality Appraisal

An analysis of study quality was conducted for each study using the Joanna Briggs Institute critical appraisal tools (Joanna Briggs Institute, 2018). These tools were designed to assess the rigor of quantitative research designs (e.g., cross-sectional and randomized clinical trials). Each included study was read completely a second time for the purpose of evaluating quality using a critical appraisal tool consistent with the reported study design. Specifically, quality was appraised for inclusion and exclusion criteria, sample adequacy, setting, validity and reliability of study methods, identification and management of potential study bias, and appropriateness of the statistical analysis plan. Based on the appraisal, it was determined that all 33 studies were of sufficient quality to be included in this analysis.

Data Extraction

Data extraction included the following broad categories: chronic disease diagnosis/diagnoses, research methods, theoretical models, study setting, sample characteristics, sampling methods, measurement tools used, factors assessed in each study, results of analysis, study limitations, professions included in the research team, and

publication information. Each of these items were recorded in a spreadsheet to aid in later analysis. Missing categories of data were noted in the spreadsheet.

Analysis Process

After data had been extracted to a spreadsheet, each category was analyzed across studies looking for patterns. Factors identified as potentially influential in transition readiness were further reviewed to determine if there was a statistically significant association to transition readiness. Those that were statistically significant were further categorized as modifiable or non-modifiable. Finally, factors found to be associated with transition readiness were grouped by similarities and patterns among the factors. In addition, gaps in the findings were noted.

Results

There were 33 studies published between 2009 and 2019 included in this analysis that assessed factors associated with transition readiness in adolescents and young adults living with a chronic disease. Articles published between 1990 and 2009 yielded no factors associated with transition readiness; rather, these articles focused on defining transition readiness and developing measurement tools to assess transition readiness. Thirty-two of the 33 studies were cross-sectional survey designs, and the remaining one was a longitudinal study. Study participant ages ranged from 12–26 years, but age ranged varied within studies. Twenty-four of 33 studies were conducted in the outpatient setting. There were four categories of factors associated with transition readiness: (a) demographic and ecological factors, (b) psychosocial factors, (c) self-management/transition readiness education factors, and (d) health outcome factors. Within each of the four categories, there were modifiable and non-modifiable categories.

Six studies used a theory or model to guide the research, including the Social-ecological Model of Adolescent and Young Adult Readiness to Transition (SMART), the Lerner Model, the Developmental Transitional Model, and the Health Care Transition Research Consortium Model.

Methodological Quality

Table 1 provides details about the findings from the quality assessment. When assessing methodological quality using the Joanna Briggs Institute critical appraisal tools, it was determined that all 33 included articles provided sufficient detail regarding the studies' inclusion and exclusion criteria, sample adequacy, setting characteristics, and appropriateness of the statistical analysis plan. The two areas that were noted to be less rigorous were use of valid and reliable measures and bias.

There were a variety of validated tools used across the studies to assess transition readiness. The most commonly utilized tools included the University of North Carolina STARx scale (Cohen et al., 2015; Ferris et al., 2015), University of North Carolina TRxNSITION (Ferris et al., 2012), and the Transition Readiness Assessment Questionnaire (Sawicki et al., 2011; Wood et al., 2014). Twenty-nine of the 33 studies utilized measurement tools that were not specific to any diseases, whereas four of the 33 studies utilized a disease-specific tool. Though a majority of the included studies utilized validated tools, six studies used non-validated tools.

The majority of the studies did not adequately address threats of bias. All 33 studies recognized the potential issues with utilizing self-reported measures of transition readiness and only measuring the adolescents and young adults' perspective; only a few studies reported using strategies to address this bias (Eluri et al., 2017; Fredericks et al.,

2010; Gilleland, Amaral, Mee, & Blount, 2011; Gilleland Marchak, Reed-Knight, Amaral, Mee, & Blount, 2015; Sawicki, Kelemen, & Weitzman, 2014; Speller-Brown et al., 2015). Six of the 33 studies included both adolescent/young adult and parent self-report as a mechanism to reduce self-report bias. A single study included a provider report of transition readiness (Gilleland Marchak et al., 2015).

Demographic and Ecological Factors

There were a variety of demographic and ecological factors found to have a relationship to transition readiness. See Table 2 for a list of demographic and ecological factors that demonstrated associations with transition readiness. Twenty-six studies assessed age, and 19 found it to be positively related to transition readiness (Beal et al., 2016; Bingham, Scalzi, Groh, Boehmer, & Banks, 2015; Dwyer-Matzky, Blatt, Asselin, & Wood, 2018; Eaton et al., 2017; Fredericks et al., 2010; Gilleland et al., 2012; Gray et al., 2015; Gumidyala et al., 2018; Haarbauer-Krupa et al., 2019; Hart, Pollock, Hill, & Maslow, 2017; Javalkar, Fenton, Cohen, & Ferris, 2014; Javalkar et al., 2016; Lazaroff, Meara, Tompkins, Peters, & Ardoin, 2018; Mackie et al., 2016; Rosen et al., 2016; Sawicki, Kelemen, & Weitzman, 2014; Speller-Brown et al., 2015; Stewart et al., 2017; Whitfield, Fredericks, Eder, Shpeen, & Adler, 2015). Studies found that increasing age was positively related to decreasing knowledge deficits in youth with sickle cell disease or heart disease (McPherson, Thaniel, & Minniti, 2009; Uzark et al., 2015). Two studies found that age was associated with increased levels of self-management skills needed for successful transition for youth with sickle cell disease or heart disease (Treadwell et al., 2016; Uzark et al., 2015) and with decreased problems with transition in the sickle cell disease population (McPherson et al., 2009). Not all studies found that

transition readiness increased with age. Four studies did not find a significant relationship between age and transition readiness in youth with inflammatory bowel disease (Carlsen et al., 2017), eosinophilic esophagitis/eosinophilic gastroenteritis (Eluri et al., 2017), kidney transplants (Fenton et al., 2015), or neurogenic bladders (Grimsby et al., 2016).

Seven studies found that female youths with a variety of chronic diseases had statistically significantly higher transition readiness scores (Eaton et al., 2017; Gray et al., 2015; Hart et al., 2017; Javalkar et al., 2014, 2016; Lazaroff et al., 2018; Sawicki et al., 2014). Additionally, females with sickle cell disease or heart disease had higher self-management behavior scores, indicating they were more prepared for transition to adult care (McPherson et al., 2009; Stewart et al., 2017). One study found that females with sickle cell disease perceived higher levels of difficulty with transition to adult care (McPherson et al., 2009). This finding about female gender was not consistent across all studies; 11 studies which assessed gender did not find an association between gender and transition readiness (Beal et al., 2016; Dwyer-Matzky et al., 2018; Eluri et al., 2017; Fenton et al., 2015; Fredericks et al., 2010; Grimsby et al., 2016; Gumidyala et al., 2018; Mackie et al., 2016; Speller-Brown et al., 2015; Treadwell et al., 2016; Uzark et al., 2015).

A few studies assessed social determinants of health factors for potential relationships with transition readiness. Positive associations for transition readiness included higher median household income (Javalkar et al., 2016), two-parent households (Beal et al., 2016; Javalkar et al., 2016), and private insurance status (Javalkar et al., 2014). Adolescents and young adults' higher health literacy (Beal et al., 2016; Bingham et al., 2015) and academic performance (Haarbauer-Krupa et al., 2019) had increased

transition readiness scores. Findings regarding the relationship between income and transition readiness was not consistent across studies. Two studies found that income did not have a relationship with transition readiness (Speller-Brown et al., 2015; Stewart et al., 2017). Other demographic and ecological factors that did not demonstrate any significant associations with transition readiness included race/ethnicity (Beal et al., 2016; Dwyer-Matzky et al., 2018; Grimsby et al., 2016; Gumidyala et al., 2018; Javalkar et al., 2016; Treadwell et al., 2016; Uzark et al., 2015), parental age (Bingham et al., 2015), parent education level (Beal et al., 2016; Bingham et al., 2015; Javalkar et al., 2014; Rosen et al., 2016; Speller-Brown et al., 2015; Stewart et al., 2017), having a job (Bingham et al., 2015; Treadwell et al., 2016), youth education level (Bingham et al., 2015; Treadwell et al., 2016), household size (Javalkar et al., 2016), parent marital status (Speller-Brown et al., 2015), and socioeconomic status (Carlsen et al., 2017; Hart et al., 2017; Rosen et al., 2016).

Disease Factors

Few associations between disease factors and transition readiness were identified. One study assessing multiple chronic diseases found that chronic conditions impacting cognitive ability were associated with lower transition readiness compared to healthy peers (Beal et al., 2016). Having a family member with a similar disease and length of illness duration were found to be positively associated with transition readiness in one study (Bingham et al., 2015). However, in other studies assessing illness duration, no relationship to transition readiness was identified (Carlsen et al., 2017; Gray et al., 2015; Javalkar et al., 2014; Pakdeeprom, In-Iw, Chintanadilok, Wichiencharoen, & Manaboriboon, 2012; Whitfield et al., 2015). In the sickle cell disease population, disease

severity was negatively associated with adolescents and young adults' interest in learning about the transition process (McPherson et al., 2009). This was not supported by other studies assessing transition readiness in adolescents and young adults living with sickle cell disease (McPherson et al., 2009; Speller-Brown et al., 2015), chronic kidney disease (Fenton et al., 2015), and inflammatory bowel disease (Gray et al., 2015; Gumidyala et al., 2018). For adolescents and young adults with congenital heart disease, having a history of a primary repair or undergoing cardiac interventions was positively associated with higher disease self-management scores and better transition readiness (Stewart et al., 2017). This same study noted the use of cardiac medications was associated with higher self-advocacy, but not with overall transition readiness (Stewart et al., 2017).

None of the studies identified an association between transition readiness and adherence to medical therapies for chronic diseases (Rosen et al., 2016; Whitfield et al., 2015). Other disease factors that did not demonstrate any significant associations with transition readiness included diagnosis (Bingham et al., 2015; Carlsen et al., 2017; Gumidyala et al., 2018; Mackie et al., 2016; Rosen et al., 2016; Stewart et al., 2017; Uzark et al., 2015), active symptoms (Eluri et al., 2017), active steroid use (Eluri et al., 2017), dietary therapy (Eluri et al., 2017), disease burden (Fenton et al., 2015), pain scores (Bingham et al., 2015), hospitalizations (Rosen et al., 2016), ambulation status (Grimsby et al., 2016), use of individualized education plan (Javalkar et al., 2014), and disease-related absences from school (Carlsen et al., 2017).

Psychosocial Factors

A variety of disparate psychosocial factors were found to be associated with transition readiness. See Table 3 for a list of psychosocial factors that demonstrated

associations with transition readiness. Self-efficacy was found to be positively associated with transition readiness in nine out of ten studies that focused on a variety of different chronic diseases and populations (Carlsen et al., 2017; Eaton et al., 2017; Gilleland Marchak et al., 2015; Gumidyala et al., 2018; Pakdeeprom et al., 2012; Sawicki et al., 2014; Stewart et al., 2017; Treadwell et al., 2016; Uzark et al., 2015). Eluri et al. (2017) was the only study that did not find a relationship between self-efficacy and transition readiness. A number of other factors associated with transition readiness were identified in single studies that used heterogeneous populations including autonomous motivation (Dwyer-Matzky et al., 2018), family cohesion (Fenton et al., 2015), social support (Pakdeeprom et al., 2012), hopeful future expectations (Hart et al., 2017), preferred source of information (Johnson et al., 2015), locus of control (Nazareth et al., 2016), patient activation levels (Lazaroff et al., 2018), anxiety (Pakdeeprom et al., 2012), disease-associated stress (Uzark et al., 2015), executive functioning (Gutierrez-Colina et al., 2017), and quality of life (Treadwell et al., 2016). Psychosocial factors that did not demonstrate any significant associations with transition readiness included overall well-being (Bingham et al., 2015), resiliency (Carlsen et al., 2017), perceived competence (Dwyer-Matzky et al., 2018), quality of life (Fenton et al., 2015; Uzark et al., 2015), coping strategies (Fenton et al., 2015), confidence in readiness to transfer (Beal et al., 2016; Gray et al., 2015), and perception of illness uncertainty (Stewart et al., 2017).

Self-Management and Transition Education Factors

There were a number of studies that identified factors related to self-management and transition education that influenced transition readiness. See Table 4 for a list of self-management and transition education factors that demonstrated associations with

transition readiness. Disease knowledge was positively related to transition readiness in studies of adolescents and young adults with congenital heart disease, kidney transplants, and other chronic diseases (Dwyer-Matzky et al., 2018; Gilleland et al., 2011; Gilleland Marchak et al., 2015; Stewart et al., 2017). Adolescents and young adults across multiple different chronic diseases who perceived greater overall responsibility for their disease self-management also had higher levels of transition readiness (Eaton et al., 2017; Gilleland et al., 2011; Gilleland Marchak et al., 2015; Haarbauer-Krupa et al., 2019; Speller-Brown et al., 2015). In addition, youth with sickle cell disease who thought they were ready to assume complete responsibility also reported higher levels of transition readiness (Speller-Brown et al., 2015).

Proactive discussions about the transition to adult care between the adolescents or young adults, parents, and providers was found to positively impact transition readiness (Gumidyala et al., 2018). Parental involvement in care was inversely associated with transition readiness for youth with a history of a kidney transplant or sickle cell disease (Gilleland et al., 2011; Gilleland et al., 2015; Speller-Brown et al., 2015). Perceived barriers to medication adherence were negatively associated with transition readiness across multiple chronic diseases (Gilleland et al., 2011; Haarbauer-Krupa et al., 2019). Finally, parents' perception of the adolescents or young adults' transition readiness was positively associated with the adolescents and young adults' overall responsibility for their disease self-management (Speller-Brown et al., 2015). Other self-management and transition education factors that did not demonstrate any significant associations with transition readiness included occurrence of transition (Jensen et al., 2017), adherence

(Fredericks et al., 2010; Rosen et al., 2016), and engagement with providers after transition (Szalda et al., 2017).

Health Outcome Factors

Only one study identified a relationship between transition readiness, and health and health care utilization outcomes. As transition readiness increased in adolescents and young adults who had undergone kidney transplants, emergency room utilization decreased (Fenton et al., 2015). In addition, transition readiness was positively related to adherence in the same study (Fenton et al., 2015). No studies assessed the relationship between transition readiness levels prior to transition, and health or health care utilization outcomes after transition.

Discussion

The purpose of this systematic review was to explore the literature regarding factors associated with transition readiness in adolescents and young adults with chronic disease, and to identify factors useful to intervention design. Thirty-three studies met the inclusion criteria and were included in the analysis. A variety of modifiable factors were found to be significantly associated with transition readiness; however, most of these factors were only studied in a single study or in a single chronic disease population. One of the most commonly identified modifiable factor was self-efficacy, which was found to be significantly, positively associated with transition readiness across multiple studies and disease populations. This finding is supported by the literature on pediatric chronic disease management where self-efficacy has been found to have a positive effect on chronic disease self-management (Noser, Huffhines, Clements, & Patton, 2017; Rhee, Belyea, Ciurzynski, & Brasch, 2009; Smith et al., 2018). Self-efficacy is an important

concept to consider when designing transition readiness interventions, because it encompasses disease self-management and the knowledge and skills needed to independently navigate the health care system.

Adolescent and young adult's responsibility for disease management, disease-specific knowledge, perceived barriers to medication adherence, and parental involvement in disease management were also found to be significantly associated with transition readiness. While these factors were not studied as frequently as self-efficacy, they show promise as factors that are potentially amenable to intervention. Adolescent and young adult's responsibility for disease management and parental involvement in disease management, otherwise known as allocation of treatment responsibility, have been studied within the context of self-management and treatment adherence, but not for transition readiness (Pai et al., 2010; Pai et al., 2011). Additionally, few studies in this review assessed concepts related to family functioning. There is a need for more research in this area as it is critical to future interventions that the family's role in development of transition readiness be accounted for. The majority of disease management occurs within the home, and it is negotiated between parents and adolescents or young adults.

Understanding how disease management is shared or distributed between parents and adolescents or young adults could help predict an adolescent or young adult's readiness to transition. It could help researchers and health care providers better understand how to develop interventions that encourage the transfer of responsibilities for disease management from parents to adolescents and young adults.

This analysis looked at primary studies that assessed adolescents and young adults with chronic conditions that encompassed a number of different diseases. This approach

was not intended to overlook the particular nuances of a specific chronic disease; rather, this strategy offered an opportunity to understand how researchers could translate current knowledge of transition readiness to other chronic disease populations. While self-management may vary by disease, knowledge found in this review of transition readiness could be applied to chronic diseases that share similar characteristics. Factors associated with transition readiness that were only studied in a single study or a single chronic disease population, but should be considered important for future research include autonomous motivation, family cohesion, social support, hopeful future expectations, patient activation levels, anxiety, and disease-associated stress. This review highlighted the need to understand more about how these modifiable factors relate to each other, and to transition readiness across different chronic disease populations.

A variety of non-modifiable factors were found to be significantly associated with transition readiness. As with the modifiable factors, many of the non-modifiable factors were studied in a single cross-sectional study or in a single chronic disease population. Age and gender were the only non-modifiable factors whose association with transition readiness was reaffirmed across multiple studies and populations. While these factors are not modifiable, they potentially impact transition readiness; therefore, it is important that researchers and health care providers understand how these factors relate to transition readiness and consider them when designing interventions. For healthy adolescents, the development of individuality and independence is often gradual, and it is guided by a progressive shift of responsibility from the parent to the adolescent. Disease self-management for an adolescent or young adult with a chronic disease would optimally take a similar approach (Michaud, Suris, & Viner, 2004; Suris et al., 2004). Researchers

should consider controlling for non-modifiable factors when designing interventions to improve transition readiness.

Other major findings of this review were gaps related to methodology. Transition readiness measurement was inconsistent across studies, and there was no agreement about the best method of measurement (de Silva & Fishman, 2014). The majority of studies included in this review used validated tools to measure transition readiness, but these tools were not disease specific. This review notes contradictory information about the benefit of disease-neutral versus disease-specific transition readiness assessment tools. It has been noted that disease-neutral tools may be the most desirable as the tools can be used to design general transition interventions that can be used across disease populations (Stinson et al., 2014). This view is disputed by Mackie et al. (2015) who noted that disease-neutral tools did not necessarily have questions that allowed them to adequately assess the adolescents and young adults' disease self-management practices specific to their disease. The benefits of disease-neutral versus disease-specific transition readiness tools need to be further explored. While some aspects of transition are the same for all adolescents and young adults, such as the change in health provider, many differences exist such as the specifics of self-management for each chronic disease. A disease-neutral tool could be used if it included subcomponents that could be tailored to the specifics of the chronic disease.

This review primarily focused on transition readiness from the perspective of the adolescent and young adult; however, transition readiness does not occur in isolation from caregivers/parents and health care providers. All three perspectives are important to developing transition readiness preparation interventions as each stakeholder has a

different perspective and role to play during this change. Future research should consider measuring transition readiness from all three perspectives so as to obtain a broader understanding of the adolescent or young adult's progress towards developing optimal transition readiness.

This review also highlighted a gap in the understanding of how transition readiness may predict health outcomes of adolescents and young adults with chronic disease after transition. Only one study assessed health outcomes in relationship to transition readiness (Fenton et al., 2015). In addition, there were no longitudinal studies to assess how transition readiness scores affected the transition experience and subsequent health outcomes after transition. Measurement tools to assess transition readiness were designed so that health care providers could assess how the adolescent or young adult was progressing towards transition readiness prior to transition (Ferris et al., 2012; Gilleland et al., 2012; Sawicki et al., 2011; Stinson et al., 2014); however, to date, no studies have utilized these tools over time to understand transition readiness from a longitudinal perspective.

Finally, the findings of this review are limited by quality and level of evidence provided in the primary studies. A majority of the included studies were cross-sectional, correlational studies using small sample sizes and lacking power analysis. Transition readiness was assessed entirely using self-report, which means that the true level of transition readiness may have been under- or over-reported. Only a few studies utilized other perspectives, such as parents or provider's views, to assess transition readiness. In addition, most of the studies were sampled from a single institution, which decreased the

ability to generalize the findings. Finally, the lack of theory supporting the studies made it difficult to align findings across the body of work.

The transition to the adult health care system has the potential for significant negative effects on the health and well-being of adolescents and young adults living with chronic disease. Transition readiness is an important area of research still requiring further descriptive research to understand factors that contribute to the development of transition readiness. Modifiable factors, such as self-efficacy, the adolescent and young adult's responsibility for disease management, disease-specific knowledge, perceived barriers to medication adherence, and parental involvement in disease management, are all amenable to intervention. Non-modifiable factors, such as age and gender, should be considered when developing research or carrying out statistical analysis. Further research is necessary to address gaps identified in this review, and there is a need for more longitudinal studies designed to provide perspective on how transition readiness changes over time and how health care providers can intervene to ensure successful transition to the adult health care setting.

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Figure 1

PRISMA Flow Diagram for the Systematic Review

Table 1 *Quality Assessment of Methodology for Included Studies*

Study	Criteria for Inclusion Defined	Study Subjects and Settings Description	Valid and Reliable Measurement	Potential Biases Identified	Used Strategies to Address Bias	Appropriate Statistical Analysis
Beal (2016)	✓	✓	✓	✓		✓
Bingham (2015)	✓	✓		✓		✓
Carlsen (2017)	✓	✓	✓	✓		✓
Dwyer-Matzky (2018)	✓	✓		✓		✓
Eaton (2017)	✓	✓	✓	✓		✓
Eluri (2017)	✓	✓	✓	✓	✓	✓
Fenton (2015)	✓	✓	✓	✓		✓
Fredericks (2010)	✓	✓		✓	✓	✓
Gilleland (2011)	✓	✓	✓	✓	✓	✓
Gilleland Marchak (2015)	✓	✓	✓	✓	✓	✓
Gray (2015)	✓	✓	✓	✓		✓
Grimsby (2016)	✓	✓	✓	✓		✓
Gumidyala (2018)	✓	✓	✓	✓		✓
Gutierrez-Colina (2017)	✓	✓	✓	✓		✓
Haarbauer-Krupa (2019)	✓	✓	✓	✓		✓
Hart (2017)	✓	✓	✓	✓		✓
Javalkar (2014)	✓	✓	✓	✓		✓
Javalkar (2016)	✓	✓	✓	✓		✓
Jensen (2017)	✓	✓	✓	✓		✓
Johnson (2015)	✓	✓	✓	✓		✓
Lazaroff (2018)	✓	✓	✓	✓		✓
Mackie (2016)	✓	✓	✓	✓		✓
McPherson (2009)	✓	✓		✓		✓
Nazareth (2016)	✓	✓	✓	✓		✓
Pakdeeprom (2012)	✓	✓		✓		✓
Rosen (2016)	✓	✓	✓	✓		✓
Sawicki et al. (2014)	✓	✓	✓	✓		✓
Speller-Brown (2015)	✓	✓	✓	✓	✓	✓
Stewart (2017)	✓	✓	✓	✓	✓	✓
Szalda (2017)	✓	✓	✓	✓		✓
Treadwell (2016)	✓	✓	✓	✓		✓
(2015)	✓	✓	✓	✓		✓
Whitfield (2015)	✓	✓		✓		✓

✓ = authors adequately addressed

Table 2 Demographic and Ecological Factors Found to have Significant or Inconsistent Relationships to Transition Readiness

Study	Chronic Disease of Focus	Age	Gender	Median Household Income	Sex Composition of Community	Household Structure	Insurance Type	Health Literacy	Academic Performance
Beal (2016)	MCD*	✓	--			✓		✓	
Bingham (2015)	Rheumatology Diseases	✓						✓	
Carlsen (2017)	IBD**	--							
Dwyer-Matzky (2018)	MCD	✓	--				--		
Eaton (2017)	MCD	✓	✓						
Eluri (2017)	Eosinophili esophagitis/ eosinophilic gastroenteritis	--	--						
Fenton (2015)	Kidney transplant	--	--						
Fredericks (2010)	Liver transplant	✓	--						
Gilleland (2011)	Kidney Transplant								
Gilleland Marchak (2015)	Kidney Transplant	✓							
Gray (2015)	IBD	✓	✓						
Grimsby (2016)	Neurogenic bladder	--	--						
Gumidyala (2018)	IBD	✓	--						
Gutierrez-Colina (2017)	Pediatric Transplant								
Haarbauer-Krupa (2019)	MCD	✓							✓
Hart (2017)	MCD	✓	✓						
Javalkar (2014)	Chronic kidney disease	✓	✓				✓		
Javalkar (2016)	MCD	✓	✓	✓	✓				
Jensen (2017)	MCD								
Johnson (2015)	MCD								
Lazaroff (2018)	Juvenile Idiopathic Arthritis	✓	✓						
Mackie (2016)	Heart disease/ transplant	✓	--						
McPherson (2009)	Sickle cell disease	✓							
Nazareth (2016)	MCD								
Pakdeeprom (2012)	MCD								
Rosen (2016)	IBD	✓							
Sawicki et al. (2014)	MCD	✓	✓						
Speller-Brown (2015)	Sickle cell disease	✓	--	--					
Stewart (2017)	Heart disease	✓		--					
Szalda (2017)	Cancer								
Treadwell (2016)	Sickle cell disease	✓	--				--		
Uzark (2015)	Heart disease	✓	--						
Whitfield (2015)	IBD	✓							

*MCD – Multiple chronic diseases, **IBD – Inflammatory bowel disease, ✓ = significance at the 0.05 level, -- = studied but no association found

Table 3 Psychosocial Factors Found to have Significant or Inconsistent Relationships to Transition Readiness

Study	Chronic Disease of Focus	Self-efficacy	Autonomous Motivation	Family Cohesion	Social Support	Hopeful Future Expectations	Locus of Control	Patient Activation Levels	Anxiety	Executive Functioning
Beal (2016)	MCD									
Bingham (2015)	Rheumatology Diseases									
Carlsen (2017)	IBD**	✓								
Dwyer-Matzky (2018)	MCD		✓							
Eaton (2017)	MCD	✓								
Eluri (2017)	Esophagitis/ Gastroenteritis	--								
Fenton (2015)	Kidney transplant			✓						
Fredericks (2010)	Liver transplant									
Gilleland (2011)	Kidney Transplant									
Gilleland Marchak (2015)	Kidney Transplant	✓								
Gray (2015)	IBD									
Grimsby (2016)	Neurogenic bladder									
Gumidyala (2018)	IBD	✓								
Gutierrez-Colina (2017)	Pediatric Transplant									✓
Haarbauer-Krupa (2019)	MCD									
Hart (2017)	MCD					✓				
Javalkar (2014)	Chronic kidney disease									
Javalkar (2016)	MCD									
Jensen (2017)	MCD									
Johnson (2015)	MCD									
Lazaroff (2018)	Juvenile Idiopathic Arthritis							✓		
Mackie (2016)	Heart disease/ transplant									
McPherson (2009)	Sickle cell disease									
Nazareth (2016)	MCD						✓			
Pakdeeprom (2012)	MCD	✓			✓				✓	
Rosen (2016)	IBD									
Sawicki et al. (2014)	MCD	✓								
Speller-Brown (2015)	Sickle cell disease									
Stewart (2017)	Heart disease	✓								
Szalda (2017)	Cancer									
Treadwell (2016)	Sickle cell disease	✓								
Uzark (2015)	Heart disease	✓								
Whitfield (2015)	IBD									

*MCD – Multiple chronic diseases, **IBD – Inflammatory bowel disease, ✓ = significance at the 0.05 level, -- = studied but no association found

Table 4 Self-Management and Transition Education Factors Found to have Significant or Inconsistent Relationships to Transition Readiness

Study	Chronic Disease of Focus	Disease-specific Knowledge	Adolescent Disease Responsibility	Discussions about Transition	Parent Involvement	Perceived Medication Barriers
Beal (2016)	MCD*					
Bingham (2015)	Rheumatology Diseases					
Carlsen (2017)	IBD**					
Dwyer-Matzky (2018)	MCD	✓				
Eaton (2017)	MCD		✓		--	
Eluri (2017)	Esophagitis/ Gastroenteritis					
Fenton (2015)	Kidney transplant					
Fredericks (2010)	Liver transplant					
Gilleland (2011)	Kidney Transplant	✓	✓		✓	✓
Gilleland Marchak (2015)	Kidney Transplant	✓	✓		✓	
Gray (2015)	IBD					
Grimsby (2016)	Neurogenic bladder					
Gumidyala (2018)	IBD			✓		
Gutierrez-Colina (2017)	Pediatric Transplant					
Haarbauer-Krupa (2019)	MCD		✓			✓
Hart (2017)	MCD					
Javalkar (2014)	Chronic kidney disease					
Javalkar (2016)	MCD					
Jensen (2017)	MCD					
Johnson (2015)	MCD					
Lazaroff (2018)	Juvenile Idiopathic Arthritis					
Mackie (2016)	Heart disease/ transplant					
McPherson (2009)	Sickle cell disease					
Nazareth (2016)	MCD					
Pakdeeprom (2012)	MCD					
Rosen (2016)	IBD					
Sawicki et al. (2014)	MCD					
Speller-Brown (2015)	Sickle cell disease		✓	--	✓	
Stewart (2017)	Heart disease	✓		✓		
Szalda (2017)	Cancer					
Treadwell (2016)	Sickle cell disease					
Uzark (2015)	Heart disease					
Whitfield (2015)	IBD					

*MCD – Multiple chronic diseases, **IBD – Inflammatory bowel disease, ✓ = significance at the 0.05 level, -- = studied but no association found

CHAPTER THREE

YOUTHS' EXPERIENCES OF TRANSITION FROM PEDIATRIC TO ADULT CARE: AN UPDATED QUALITATIVE METASYNTHESIS

Varty, M., Speller-Brown, B., Phillips, L., & Kelly, K.P. Youths' experiences of transition from pediatric to adult care: An updated qualitative metasynthesis. *Journal of Pediatric Nursing*. Manuscript is currently in review.

Abstract

Problem: Improvements in chronic disease management has led to increasing numbers of youth transitioning to adult healthcare. Poor transition can lead to high risks of morbidity and mortality. Understanding adolescents and young adults perspectives on transition is essential to developing effective transition preparation. The aim of this metasynthesis was to synthesize qualitative studies assessing the experiences and expectations of transition to adult healthcare settings in adolescent and young adults (AYA) with chronic diseases to update work completed in a prior metasynthesis by Fegran and colleagues (2014).

Eligibility criteria: A search of PubMed, Medline, PsycINFO, and CINAHL was conducted to gather articles published after February 2011.

Sample: Of 889 articles screened, a total of 33 articles were included in the final analysis.

Results: Seven main themes were found including: developing transition readiness, conceiving expectations based upon pediatric health, transitioning leads to an evolving parent role, transitioning leads to an evolving youth's role, identifying barriers due to differences between pediatric and adult healthcare settings, lacking transition readiness leads to poor results, and recommendations for improvements to transition preparation.

Conclusions: Findings of this metasynthesis reaffirmed previous findings and highlighted the need to create adolescent and young adult-centered transition preparation.

Implications: Transition preparation interventions need to be developed which recognize the ongoing changes in AYA development, changes in AYA and parent roles, and the need to prepare AYA for the differences between the pediatric and adult healthcare setting.

Keywords: adolescents and young adults, transition, transition readiness, qualitative metasynthesis

With increasing numbers of youth with chronic diseases living into adulthood, preparation for transition to adult healthcare settings merits attention. Though transition has been a recognized issue since the 1980's, transition preparation and interventions are not consistently provided when appropriate, and health outcomes after transfer demonstrate higher risks of morbidity and mortality for young adults (Blinder et al., 2013; Blum et al., 1993; Dickerson, Klima, Rhodes, & O'Brien, 2012; McManus et al., 2013; Quinn, Rogers, McCavit, & Buchanan, 2010). Transition has been described as a complex process rather than just a handoff of the youth's care from the pediatric healthcare team to the adult healthcare team. It involves, but is not limited to, transfer of disease management from parents to the adolescent or young adults (AYA), coordination of a new healthcare team to take over responsibility for the AYA, and transfer of appropriate medical records and information necessary for continuity of care (Meleis, 2010).

Many recommendations have been made to encourage the development of transition interventions to prepare AYAs for transition; however, AYAs still face barriers during this time period (Fegran, Hall, Uhrenfeldt, Aagaard, & Ludvigsen, 2014; McManus et al., 2013). Pre-transition barriers include the AYA's negative feelings towards transfer to an adult healthcare setting (de Silva & Fishman, 2014; Wong et al., 2010), inadequate self-management skills and disease knowledge (de Silva & Fishman, 2014; Gilliam et al., 2011) and AYA's lack of interest in leaving their comfortable pediatric environment (Dowshen & D'Angelo, 2011; Rutishauser, Sawyer, & Ambresin, 2014). Barriers to establishing care in the adult healthcare setting include lack of referral to a healthcare provider (Garvey et al., 2014), poor communication between the pediatric

and adult healthcare providers (de Silva & Fishman, 2014; Garvey et al., 2014; Wong et al., 2010), and not being offered the chance to have a clinic visit with the adult healthcare provider before being discharged from pediatric healthcare (Garvey et al., 2014). After transition, AYAs face challenges including inconsistency between the pediatric and adult healthcare settings (Goossens et al., 2011; Park, Adams, & Irwin, 2011). Poor health outcomes seen after transition such as poor clinic attendance, poor medication adherence, and even complete cessation of all medical care have been attributed to poor transition practices (de Silva & Fishman, 2014; van Staa, van der Stege, Jedeloo, Moll, & Hilberink, 2011). Developing interventions for preparation and to improve the handoff between the different healthcare settings is essential to reducing these difficulties faced by AYAs during the transition time period.

Understanding the AYAs' perspectives on transition is instrumental to developing effective interventions which will be perceived as relevant and informative. Fegran et al. (2014) completed a metasynthesis on qualitative studies assessing AYAs' experiences of transition from 1999 through February 2011. Given the increasing attention to transition care and an expanding qualitative evidence base, including seeking the AYAs' perspectives, this metasynthesis was conducted to update the work completed by Fegran et al. (2014). This metasynthesis looked to synthesize more recent articles and to build upon previous findings. The research questions which guided this metasynthesis were: a) What are AYAs' perspectives of transition preparation and transfer to the adult healthcare setting? b) What are their recommendations for improving the transition process?, and c) How do these findings compare to the findings of the prior metasynthesis?

Aims

The aim of this metasynthesis was to update and compare prior work completed by Fegran and colleagues by synthesizing qualitative studies published since 2011 assessing the experiences and expectations of transition to adult healthcare settings in AYA with chronic diseases.

Methods

The methods for this metasynthesis were designed using Sandelowski and Barroso's (2007) guidelines for synthesizing qualitative research (Sandelowski & Barroso, 2007).

Inclusion Criteria

In order to build upon work completed by the prior metasynthesis, similar selection criteria were chosen to identify relevant literature. For a study to be included it needed to comprise (or contain) either adolescent's or young adult's experiences or expectations with transition preparation and transfer from the pediatric to the adult healthcare setting. AYAs in the study must have been diagnosed with a chronic disease. The study also needed to incorporate a qualitative design and to be peer-reviewed and published in English.

Search Strategies

A systematic search was conducted in PubMed, Medline, PsycINFO, and CINAHL in October 2018 with supplementary searches conducted in May 2019 and October 2019 to capture relevant articles which were newly published. The search strategy included the key words "youth OR teenager OR young adult OR adolescent" AND "sickle cell OR epilepsy OR diabetes OR cystic fibrosis OR chronic kidney disease OR cerebral palsy OR neurogenic bladder OR spina bifida OR congenital heart disease

OR asthma OR HIV OR transplant OR chronic disease OR chronic illness” AND “transfer OR transition” AND “qualitative study OR phenomenology OR grounded theory OR hermeneutics OR qualitative”. The prior metasynthesis in this area assessed literature from 1999 through February 2011. This metasynthesis sought to identify articles published March 2011 through January 2019. In addition bibliographies of relevant articles were searched to identify additional articles.

Search Outcome

A PRISMA diagram (Figure 2) reflects the identification, inclusion, and exclusion of articles during the search process. A total of 889 records were identified through multiple database searches. After removing duplications a total of 648 abstracts were reviewed for relevance. A total of 53 full-text articles were retrieved since the abstracts appeared relevant to addressing the aims of this metasynthesis. Full-text articles were assessed using the inclusion and exclusion criteria. In total, 32 articles were included in the final analysis.

Study Characteristics

A majority of the studies were conducted in the United States; however, the studies were also conducted in the United Kingdom, Sweden, Germany, Switzerland, Canada, Belgium, Brazil, and France. Not all studies reported gender but across those which did, 55% of the participants were female. Fifteen different chronic condition types were reported. The studies utilized individual interviews, focus groups, and qualitative questionnaires to collect data. One study utilized a mixed-methods design to collect data, but only the qualitative data were utilized for this metasynthesis. More information on the studies' characteristics can be found in Supplement 1.

Assessment of Methodological Quality

The Critical Appraisal Skills Programme (CASP) qualitative checklist was utilized to assess the quality of the studies. Two independent reviewers conducted quality appraisal (M.V. and K.P.K.). One study was excluded as result of the appraisal. Overall, a majority of the studies had clear statements of the aims, congruence between study purpose and methodology choices, appropriate research design and recruitment strategies, and clear statements of the findings. Only a few minor concerns were found regarding the rigor of data analysis. The CASP appraisal for the included articles can be found in Supplement 2.

Data Extraction and Synthesis

Descriptive data were extracted from the primary studies including: chronic condition type, study aim, method of analysis, sample size, context (country in which study was conducted), process of recruitment, gender of participants, age range, data collection process, and summary of findings. Then the primary author (M.V.) extracted all the primary study findings to be analyzed utilizing the method developed by Sandelowski and Barroso (2007). M.V. conducted a thematic analysis on the pooled extracted study findings for all 32 articles. As new themes were recognized, sub-themes were created, and as repetitive sub-themes with similar content were recognized, subthemes were collapsed into themes. This process of reviewing the primary texts, creating, and collapsing sub-themes occurred until the researcher did not recognize any new themes, and the final theme and sub-themes could no longer be collapsed any further. An audit trail was maintained using a code book. After this initial analysis, two members of the study team (B.S.B. and L.P.) independently re-analyzed the extracted

findings using the coding structure. Themes and subthemes were revised by the three primary researchers during team discussions. All disagreements were discussed until a consensus was reached.

The GRADE-CERQual approach was implemented in the analysis to provide an assessment of how much confidence could be placed in each of the theme findings.

Lewin et al. (2010) developed this assessment to provide a transparent method for evaluating the findings of qualitative synthesis. It provides a structured approach which assesses four major components of the included studies that contributed evidence to the synthesis findings. These included methodological limitations, adequacy of the data, coherence, and relevance (Lewin et al., 2015). Methodological limitations refer to the issues related to the design or way in which the study was carried out (Lewin et al., 2015). The CASP appraisal checklist was utilized to assess methodological limitations. Adequacy of the data refers to quantity and degree of richness of data supporting a finding (Lewin et al., 2015). Coherence refers to how clear and convincing the fit is between the data from the original studies and synthesized findings of the final review (Lewin et al., 2015). Finally, relevance refers to the extent to which the primary study findings supporting a synthesis finding is pertinent to the specific context (i.e. setting, population) of the metasynthesis research question (Lewin et al., 2015). More information on the GRADE-CERQual assessment for each main theme can be found in Supplement 3.

Results

AYAs' experiences of transition preparation and processes from the 32 primary studies illustrated seven main themes: a) developing transition readiness, b) conceiving

expectations based upon pediatric healthcare, c) transitioning leads to an evolving parent role, d) transitioning leads to an evolving youth role, e) identifying barriers, f) lacking transition readiness, and g) recommendations for improvements. Definitions of the main themes can be found in Table 5.

The GRADE-CERQual approach was used by the researchers to determine the confidence level for the evidence contributing to each main theme. Themes were rated as having high confidence included “Developing Transition Readiness,” “Transitioning Leads to Evolving Parent Role,” “Lacking Transition Readiness,” and “Recommendations for Improvement.” Themes rated as having moderate confidence included “Conceiving Expectations Based Upon Pediatric Healthcare,” “Transitioning Leads to Evolving Youth Role,” and “Identifying Barriers.”

Developing Transition Readiness

Increasing knowledge, especially information regarding what would happen after transition, provided comfort to AYAs, and they viewed it as a necessary tool for their new role after transition. Having information about the transition process alleviated surprise (Asp, Bratt, & Bramhagen, 2015; Lopez et al., 2015; Sobota, Umeh, & Mack, 2015). Disease knowledge assisted them to determine which healthcare providers they needed to continue to see in the adult healthcare setting (Asp et al., 2015; Lewis & Noyes, 2013; Sobota et al., 2015). It also helped them to understand why they had undergone specific examinations in the past, why their treatment was important, and how to navigate going forward (Asp et al., 2015; Porter, Graff, Lopez, & Hankins, 2014). For AYAs, knowledge was the key to understanding the healthcare delivery process but was also a mechanism to increase their role in disease management.

Practicing independence in pediatric appointments readied AYAs for their new role after transition. Upon entering the adult healthcare setting, AYAs recognized a change in how the adult provider interacted with them in comparison to their pediatric provider, and AYAs felt that pediatric providers could assist with this difference by giving them an active role earlier (Nicholas et al., 2018; Ritholz et al., 2014; Sobota et al., 2015). They felt that taking part in self-management behaviors and engaging independently with pediatric providers let them learn skills for adulthood (Asp et al., 2015; Hilliard et al., 2014; Nicholas et al., 2018; Porter et al., 2014). Not all AYAs valued this practice while in pediatric health care settings, but retrospectively they found it to be beneficial (Sobota et al., 2015).

Trying out the new adult healthcare team prior to transition was another way to decrease the burden after transition. Meeting the team prior to transition increased feelings of security and confidence (Asp et al., 2015; Hilderson et al., 2013) and decreased feelings of uncertainty (Nicholas et al., 2018; Wright, Elwell, McDonagh, Kelly, & Wray, 2016). Having an overlapping clinic with both pediatric and adult providers helped to assure AYAs that information about their condition was going to be passed along; it also facilitated the development of their relationships with their new providers (Asp et al., 2015; Hilderson et al., 2013; Morsa et al., 2018; Porter et al., 2014; Tierney et al., 2013).

Developing independence and self-management was viewed as a natural development step for some AYAs during transition. Leaving pediatrics made them feel apprehensive; however, the benefit of leaving pediatrics included gaining independent decision-making and changing their relationship style with their provider (Bemrich-Stolz,

Halanych, Howard, Hilliard, & Lebensburger, 2015; Burström, Öjmyr-Joelsson, Bratt, Lundell, & Nisell, 2016). Just as transferring settings was natural, so was the shifting of responsibilities between parents and AYAs (Hilliard et al., 2014). Providers could ensure that AYAs had increased independence by encouraging self-management, accountability, and providing education on disease self-management (Lewis & Noyes, 2013; Ritholz et al., 2014).

Conceiving Expectations Based Upon Pediatric Healthcare

Depending on when they were diagnosed with a chronic disease, AYAs may have spent years getting to know their pediatric providers which influenced their thoughts about future patient-provider relationships after transfer. They developed assumptions about their adult healthcare teams based upon their pediatric providers and worried about changes to come after transition (Wright et al., 2016). AYAs who had positive experiences with their pediatric team expected the same competence and level of familiarity with their new providers (Asp et al., 2015; Burström et al., 2016; Carroll, 2015; Morsa et al., 2018). Having strong relationships with pediatric providers led to AYAs having feelings of anxiety prior to transition (Wright et al., 2016). When they were not prepared for relationship changes, they expressed surprise with their first impressions (DiFazio, Harris, Vessey, Glader, & Shanske, 2014; Garvey et al., 2014; Huang et al., 2011; Lochridge, Wolff, Oliva, & O'Sullivan-Oliveira, 2013). For AYAs with severe disease who had been more reliant on parents for disease management, they felt they could not meet the expectations of the adult providers and could not express themselves adequately (Huang et al., 2011). Change was not always negative though, as AYAs did

see opportunity for personal growth as adult providers expected more in comparison to pediatric providers (Huang et al., 2011).

After transition, AYAs wanted to establish a trusting relationship with their adult provider. They had positive memories of their pediatric providers which influenced this desire (Carroll, 2015; Lewis & Noyes, 2013). Developing familiarity and trust with providers was considered a necessary step from the AYA's perspective to smooth transition and helped to keep them engaged in their healthcare (Lewis & Noyes, 2013; Lochridge et al., 2013; Plevinsky, Gumidyala, & Fishman, 2015). AYAs described factors that helped to build trust including continuity of care, open communication active relationship building, and attentiveness to the AYAs' needs (Lewis & Noyes, 2013; Plevinsky et al., 2015; Wright et al., 2016). Factors that hindered the development of trust were AYAs' concerns about the adult providers' competence or concern that they would change their current treatment regimen (Lewis & Noyes, 2013; Lochridge et al., 2013; Porter et al., 2014). AYAs expressed that relationship building took time but could lead to a sense of partnership (Plevinsky et al., 2015; Price et al., 2011).

After transition, AYAs expected that their new adult healthcare provider would assist them in developing self-management and tailor this support to where the AYA was developmentally. Transition is a process and not all AYA are in the same place after transfer regarding their knowledge and abilities. They expected that the team should tailor their communication and expectations to meet the individual's needs (Hilliard et al., 2014; Price et al., 2011). Providers could help AYA to self-manage by giving them practical advice regarding symptoms, lifestyle management, and self-care (Lewis & Noyes, 2013). Empowerment through assisting them to problem-solve and achieve

independence in their healthcare was seen as a necessary and beneficial task of the adult provider (Price et al., 2011).

Transitioning Leads to an Evolving Parent Role

As the AYAs were heading into an unfamiliar setting with a new healthcare team, they continued to view their parents as a resource. During transition, they were enjoying their independence; however, they still found discussing their health with parents to be reassuring (Allen, Channon, Lowes, Atwell, & Lane, 2011). They wanted their parents' role to focus less on disease management and more on providing support (Allen et al., 2011; Hilderson et al., 2013; Morsa et al., 2018). They were a resource for AYAs as they were trying to make decisions (Allen et al., 2011; Burström et al., 2017). Just maintaining connectedness with parents was considered a strength during this time (Ridosh, Braun, Roux, Bellin, & Sawin, 2011).

AYAs reported that transition required active participation from parents and reported specific functions which parents could assume in their new role during transition. Parents could advocate for them in adult healthcare, help them to remember their medical history, and help avoid misunderstandings with providers (Asp et al., 2015; Hilderson et al., 2013; Lewis & Noyes, 2013; Lochridge et al., 2013; Nicholas et al., 2018; Ritholz et al., 2014). Parents acted as back-up for youth during this time (Hilliard et al., 2014). They could offer companionship during their first visits to the adult healthcare setting (Allen et al., 2011; Asp et al., 2015; Burström et al., 2017; Hilderson et al., 2013; Lewis & Noyes, 2013; Ridosh et al., 2011; Wright et al., 2016). In addition, they aided AYAs during this time by slowly giving responsibility over to the AYA which was well-received by AYAs (Sobota et al., 2015).

Transitioning Leads to an Evolving Youth's Role

Just as parents had an evolving role during transition, AYAs also recognized that their role would be changing and they saw the value and importance in assuming independence. AYAs expressed that the expected role change was natural and developing self-management was part of growing up (Lochridge et al., 2013; Morsa et al., 2018; Nguyen et al., 2016; Nicholas et al., 2018; Porter et al., 2014; Sobota et al., 2015). AYAs viewed this change in roles as an incremental process (Asp et al., 2015; Nicholas et al., 2018); they valued gaining knowledge and skills which would assist in increasing their ability to self-manage their disease and engage with their new providers (Lewis & Noyes, 2013; Sobota et al., 2015). To move forward with their life in areas like their education or careers, they knew they had to be more independent in their healthcare (Lochridge et al., 2013). This progression towards independence was viewed in a positive light even though they recognized many barriers to navigating transition such as identifying a new healthcare provider, making appointments, and understanding insurance issues (Plevinsky et al., 2015). Healthcare providers could assist with the role change by promoting and supporting the AYAs' responsibility and accountability (Ritholz et al., 2014; Sobota et al., 2015).

During transition, AYAs expressed a desire to feel like an adult and take on the adult role. They remarked that they liked that their adult healthcare provider treated their appointments more business-like (Hilderson et al., 2013). For youth with minor disease issues, this change in approach was welcomed as they wished to rely less upon others (Ridosh et al., 2011; Ritholz et al., 2014). However, AYAs with higher disease severity did not find this efficiency to be sufficient to address their worries (Huang et al., 2011).

Transitioning to the adult healthcare system offered them an opportunity to increase independence, gain confidence in handling their medical issues, and reduced their feeling of being out of place in the pediatric setting (Lochridge et al., 2013). Being treated like an adult was also seen as a motivating factor to stay involved in their healthcare (Price et al., 2011). AYAs did also admit though that this adaption would need time for them to be comfortable (Tierney et al., 2013).

Identifying Barriers

AYAs felt that adult healthcare providers' approach made it challenging to establish a new relationship with them after transition. AYAs viewed adult healthcare providers as less personable which suggested to the AYAs that they did not want to develop a relationship (Hilliard et al., 2014). In addition, AYAs also felt like actions taken by the adult healthcare providers were sometimes biased causing the youth adult to experience poorer treatment exposing them to worse outcomes (Bemrich-Stolz et al., 2015). Specific issues regarding adult healthcare providers' approaches included their reduced time for appointments (Hilliard et al., 2014) and putting primary focus on clinical parameters during interactions (Tierney et al., 2013). Transition preparation did not necessarily lessen these concerns (Price et al., 2011). Even if the adult healthcare provider was capable or had the knowledge to care for the AYAs, the youth felt that this did not make up for the lack of personal connection (Wright et al., 2016).

AYAs expressed distress over losing their relationship with their pediatric provider at the time of transition. Pediatric providers were viewed like family and an important emotional connection (DiFazio et al., 2014; Porter et al., 2014). Having spent a period of time with their pediatric provider and having gotten used to a specific standard

of care, AYAs developed reluctance towards transitioning (Lochridge et al., 2013; Porter et al., 2014). They also expressed feelings of abandonment (DiFazio et al., 2014). The loss of this relationship coupled with a negative experience in adult healthcare system prompted the AYA youth to return to the pediatric setting to seek care (Lochridge et al., 2013).

As they were struggling to let go of their pediatric healthcare teams, AYAs also expressed feelings that their adult providers did not know as much as their pediatric providers. In their first experiences with adult providers in places such as the emergency room and primary care, they noticed that the providers offered different treatments which made them think that the adult providers lacked expertise (Bemrich-Stolz et al., 2015; Carroll, 2015; DiFazio et al., 2014). Their adult providers did not give AYAs the impression that they knew them at all (Wright et al., 2016). For example, when the adult providers asked repetitive questions during appointments about medical history or disease self-management, this reinforced that adult providers were not acquainted with them and less capable than their pediatric providers (Porter et al., 2014; Wright et al., 2016).

Lacking Transition Readiness

When AYAs were not prepared for transition it shaped their perspectives of their future healthcare team even before they left the pediatric setting. Multiple studies found that youth either only had a brief conversation about transition or transition was not mentioned at all (Asp et al., 2015; Hilderson et al., 2013; Plevinsky et al., 2015). Abrupt transition led them to feel that independence was forced upon them (Hilderson et al., 2013). They did not understand how this change in their care team would impact them, why they had to make the change, who was responsible for them, or who would be a

resource for them (Asp et al., 2015; Carroll, 2015; Catena et al., 2018; DiFazio et al., 2014; Hilderson et al., 2013; Lochridge et al., 2013; Morsa et al., 2018). They felt like novices as they interacted with adult providers as a result (Carroll, 2015). The uncertainty in their expectations was associated with reports of anxiety, fear, sadness, feelings of abandonment, and concerns about the future (Bemrich-Stolz et al., 2015; Garvey et al., 2014; Hilderson et al., 2013; Lochridge et al., 2013; Porter et al., 2014; Porter, Rupff, Hankins, Zhao, & Wesley, 2017).

AYAs reported that they did not have enough disease knowledge to actively participate in their healthcare after transition. AYAs reported not knowing specific details about their condition (Asp et al., 2015; Burström et al., 2016; Morsa et al., 2018; Ridosh et al., 2011; Sobota et al., 2015), and they reported being concerned that they did not know signs of deterioration for their conditions or when to seek help (Asp et al., 2015). They also reported uncertainty about how their disease could impact their future (Bomba, Herrmann-Garitz, Schmidt, Schmidt, & Thyen, 2017; Lopez et al., 2015). Lacking knowledge led AYAs to feel embarrassment and concern about conversations with their adult providers as they may not be able to answer all of the provider's questions (Burström et al., 2016; Tierney et al., 2013). They felt lacking knowledge could make the difference in their health outcomes (Porter et al., 2017).

Lack of preparation for change in healthcare setting was associated with AYAs reports of the new environment being discomfoting. They did not realize that they would have increased exposure to more of the long-term complications of their chronic disease or an older population (Garvey et al., 2014; Hilderson et al., 2013). This induced feelings of being in limbo. They felt too old for the pediatric setting but too young for the adult

setting (Hilderson et al., 2013). Specifically, they did not feel prepared for the different approach of the healthcare team. The new environment was less friendly, unpleasant in terms of aesthetics, was too hurried, and mainly disease-focused rather than person-centered (Catena et al., 2018; Lochridge et al., 2013; Morsa et al., 2018; Porter et al., 2017; Tierney et al., 2013). There was a decrease in the frequency of their follow-up appointments which led them to feel forgotten at times, get away with as minimal care as possible, and sometimes to disengage all together, especially if they perceived the adult provider did not care about them (Huang et al., 2011; Lewis & Noyes, 2013).

After AYAs transitioned to the adult healthcare setting, they felt uncertain of how to engage in the new setting. Though they want to be involved in their healthcare, AYAs' lack of preparation left them unable to thoroughly understand what their adult healthcare team was asking of them or the language they used (Asp et al., 2015). Working alone with their provider made this lack of preparation increasingly evident (Asp et al., 2015). Even if they had begun to become more independent in disease self-management, this did not alleviate their concerns about feeling unprepared (Tierney et al., 2013). When parents had been responsible for most of the AYAs' care, AYAs found it confusing to understand what their role and responsibilities would look like when in the adult healthcare setting (Asp et al., 2015). They reported being nervous about having conversations with adult providers as they were used to the communication styles of their pediatric providers (Wright et al., 2016). On a practical level they lacked preparation in identifying healthcare providers and other health resources (Bemrich-Stolz et al., 2015; Garvey et al., 2014; Huang et al., 2011). They found it challenging to understand insurance and how insurance affected their care (Bemrich-Stolz et al., 2015). When an AYA was not

engaged in the system or with their new adult healthcare team after transition, then they often sought care at the minimum necessary (Garvey et al., 2014; Lewis & Noyes, 2013).

Recommendations for Improvements to Transition Preparation

AYAs wanted age pertinent information in light of living with a chronic disease. They asked that knowledge be given to them which was appropriate for where they were at personally in their lives (Bomba et al., 2017; Burström et al., 2017; Burström et al., 2016; Catena et al., 2018; Garvey et al., 2014; Hilderson et al., 2013; Lewis & Noyes, 2013; Lopez et al., 2015). For example, females wanted information on pregnancy and contraception, and they wanted healthcare providers to think carefully about timing of providing information as waiting until adult healthcare was often too late for this information (Bomba et al., 2017; Burström et al., 2016; Catena et al., 2018; Lopez et al., 2015). Both genders talked about wanting to know about hereditary issues (Burström et al., 2016). Other topics which AYAs reported desiring more knowledge on included drugs, alcohol, and other lifestyle related information (Bomba et al., 2017; Burström et al., 2017; Garvey et al., 2014; Lopez et al., 2015; Morsa et al., 2018; Nicholas et al., 2018; Price et al., 2011). AYAs wanted pediatric and adult providers to recognize that emerging adults were a specific developmental group with unique needs (Bomba et al., 2017; Carroll, 2015; Freeman, Stewart, Cunningham, & Gorter, 2018; Garvey et al., 2014; Hilderson et al., 2013; Lopez et al., 2015).

AYAs also reported wanting specific details on how the transfer process would work. They recognized that they would go through a “system change” and wanted information on how this would be carried out (Bomba et al., 2017). They wanted lists of things to do in preparation (Lopez et al., 2015). Information on topics such as making

appointments, filling prescriptions, communication with providers, and identifying future healthcare providers was valued (DiFazio et al., 2014; Lochridge et al., 2013; Lopez et al., 2015; Porter et al., 2017). They expected this to be provided in the pediatric setting or through programing directly aimed at emerging young adults (Garvey et al., 2014). They felt this would appropriately shape their expectations (Garvey et al., 2014). Providing this earlier rather than later was desired (DiFazio et al., 2014; Hilliard et al., 2014).

In providing this developmental-specific knowledge, AYAs desired for their pediatric providers to talk to and address them directly to model adult healthcare. They wanted their provider to address them and not their parents as this allowed them to reflect more on themselves and their healthcare needs (Burström et al., 2017; Tierney et al., 2013). Having the opportunity to discuss their opinions made them feel like an active partner in their care (Hilderson et al., 2013). Though AYAs wanted to be directly addressed and to have their pediatric providers should move away from the “parent-centric” approach, they still expressed a desire for their parents to remain involved while they were preparing for transition (Ritholz et al., 2014).

Another common suggestion to improve transition preparation was to engage AYAs with peers who had similar experiences. They felt that meeting with their peers would help them to learn how to prepare for transition (Morsa et al., 2018; Porter et al., 2017; Wright et al., 2016). Transition was seen as a challenging time for AYAs, and peers could be a resource (Burström et al., 2017; Lochridge et al., 2013; Lopez et al., 2015; Wright et al., 2016). Peers could provide support and mentorship for life before and after transition (Bomba et al., 2017; Freeman et al., 2018; Garvey et al., 2014; Lopez et al., 2015; Morsa et al., 2018; Porter et al., 2017). Peers could assist AYAs in

developing communication skills (Burström et al., 2017). They could talk about issues that affect them daily such as friends, work, family planning, or any of the number of issues which would affect them in the future (Bomba et al., 2017; Hilderson et al., 2013). It did not matter if these peers came from similar backgrounds or had similar demographic characteristics (Lopez et al., 2015).

AYAs also spoke of the importance of coordination between pediatric and adult healthcare settings as a strategy to improve transition; they wanted the two systems to work together to ease the transfer process. AYAs felt that there should be communication between these two systems to help with continuity of care (Bomba et al., 2017; DiFazio et al., 2014; Huang et al., 2011; Nicholas et al., 2018). It was suggested that a period of overlap or having joint visits with the pediatric and adult providers could be beneficial (Bomba et al., 2017; Burström et al., 2017; Hilliard et al., 2014; Nicholas et al., 2018; Price et al., 2011). This overlap would facilitate relationship development with their future healthcare providers (Bomba et al., 2017). Meeting the team prior to transfer would help to decrease uncertainty and increase comfort with the new team (Wright et al., 2016). Finally, AYAs wanted their pediatric providers to stay connected after transition to ensure successful transfer (Ritholz et al., 2014).

To improve upon transition, AYAs talked about the importance of having a structured transition process. They wanted a formal process which introduced them to transition which offered them opportunity for learning (Bomba et al., 2017; Lochridge et al., 2013). They offered numerous suggestions for interventions to include in a structured process, including roleplaying, shadowing, providing written material, overlapping visits and engaging peers (Garvey et al., 2014; Lochridge et al., 2013; Porter et al., 2017). AYA

felt that transition processes could encourage self-efficacy (Asp et al., 2015), self-advocacy, and self-management (DiFazio et al., 2014). AYAs reported that when they did not have structured preparation then transition felt like a foreign concept (Carroll, 2015).

Discussion

Due to improvements in pediatric chronic disease management increasing numbers of AYAs are transitioning to adult care. Our update of Fegran and colleagues' metasynthesis covering qualitative research through February 2011 demonstrates increased research focus in this area. Between March 2011 and January 2019, an additional 32 articles met inclusion criteria almost doubling the previous research in this area. Our findings reaffirmed previous AYAs' transition experiences and underscored the need to create AYA-centered transition preparation now. Our findings highlight the complexity of the transition process that requires preparation of both AYA and parent across a multi-year focus of care coordination to ready the AYA for transition to adult care. Coordination begins in the pediatric setting but must continue in the adult care settings as the young adult learns the particulars of what to expect of their adult providers but also what their adult providers expect of them. In addition a complex transition of both parent and AYA roles occurs as the AYA prepares to enter the adult care medical complex.

Implications for Practice Translation of Consistent Research Findings

Comparing our synthesis with the previous work of Fegran et al. we identified numerous findings that were similar and thus are now ready for practice integration. AYAs continued to report feeling unprepared for transition and identified barriers to the process (Fegran et al., 2014). Lack of preparation led to uncertainty about the new adult

setting, AYAs were transferred based upon age and not readiness. They reported negative feelings toward transfer from pediatric adult care as they recognized differences between the pediatric and adult setting and formed assumptions about the adult healthcare team. Our work underscores the importance of starting transition preparation early and that that increasing knowledge and self-management is key to transition readiness. Self-management and independence need to begin in the pediatric setting with continuous support from the adult providers after transition. Shifting responsibilities between AYAs and parents was viewed as a natural yet valuable process, while AYAs recognized that the parents' role would evolve and change overtime. Parents moved from being primary caregivers to a supportive role; however, they still held a critical role in assisting the AYA with communication with new adult providers. Parents often remembered medical history and could help to avoid misunderstandings with providers. They could be valuable during initial visits to adult healthcare and a resource for decision-making. These are important factors to consider when developing a transition program or facilitating transition from pediatric to adult care.

Our metasynthesis also found similar reports of AYA grieving the loss of longstanding pediatric relationships (Fegran et al., 2014). Upon meeting their new adult providers, they experienced negative first encounters due to their perceived impersonal and disease-focused approach of adult healthcare (Fegran et al., 2014). Effort should be made to prepare AYA for the change in environment and for ways to self-advocate within their new medical teams. There should also be steps taken to increase AYA-centered communication and approaches within adult healthcare practices. AYAs are a specific population with unique developmental needs that need to be addressed. This is critical as

our findings demonstrate that when expectations were met, they were more likely to stay engaged with care.

The American Academy of Pediatrics (AAP), American Academy of Family Physicians (AAFP) and American College of Physicians (ACP) joint Consensus Statement on Health Care Transition for Young Adults with Special Health Care (2002) promoted an organized comprehensive coordinated method of transition that involved transition readiness assessment, preparation and planning and transition. In 2009 the Six Core Elements of Health Care Transition™ were developed to improve transition and have been promoted by Got Transition®, a cooperative agreement between the Maternal and Child Health Bureau and the National Alliance to Advance Adolescent Health (Got Transition, 2014). However almost two decades since the original consensus statement and a decade since the development of the core elements, the themes that arose in the 32 primary studies illustrated continued deficiencies in each of these six areas of transition. Table 6 provides a list of the Six Core Elements of Health Care Transition™ and links them to AYAs' reports from the metasynthesis regarding deficiencies and recommendation for practice improvements which are ready for translation.

Implications for Future Research

Research is needed to evaluate the impact of wrap around services with equal focus on pediatric and adult care facilities. Thus far the majority of this research has been conducted by pediatric providers; however, including adult providers is critical to solving the transition process for AYAs with chronic illness. Further research is needed to understand what factors may help to better facilitate relationships between AYA and their new adult healthcare providers. This could include assessing healthcare provider and staff

communication, roles of healthcare providers and staff, and clinic structure/processes. In particular, the role of the family nurse practitioner should be assessed, as they could engage in care of AYAs across the transition process and remain involved in the AYAs care as a consistent healthcare provider across the settings.

Family research should examine the evolving roles of both parent and AYA, including the impact on and care needs within the adult care environment. The most vulnerable time for the transitioning AYA is the time immediately after transfer. New findings from this metasynthesis underscore role transition of both parent and AYA in conjunction with a need for adult providers to recognize and provide additional supports for both during this initial transition timeframe. AYAs reported that parents should continue to have a role post-transition; however, they did not clearly define this role. Further research should be completed to understand these roles and how they could impact transition success.

Conclusions

This metasynthesis has demonstrated a call to arms for improvement in transition processes because AYA with chronic illness continue to report leaving pediatric care without adequate preparation. Lack of preparation influences AYAs' perspective of the future health care team. Preparation must focus on both the AYA and family. Care coordination needs to traverse the pediatric and adult care settings. Consistent themes that were essential components of successful transition include: having a structured transition process, assessment of readiness, ensuring self-management, and establishing trusting relationships with the adult health care team. These ought to receive priority attention to improve the likelihood of successful transition for AYAs living with a chronic disease.

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Figure 2

PRISMA Flow Diagram for the Metasynthesis

Table 5

Main Themes and Definitions

Main Theme	Definition
Developing transition readiness	Adolescents and young adults view transition preparation as a time to obtain certain levels of knowledge, experience, support, and independence to help them succeed after transition to the adult healthcare setting.
Conceiving expectations based upon pediatric healthcare	Over their years of being taken care of by pediatric healthcare providers, adolescents and young adults have developed assumptions about their future adult healthcare teams which they carry with them while they are preparing for transfer and after they transfer to the adult healthcare setting.
Transitioning leads to evolving parent role	During transition to adult healthcare, AYA develop independence but still desire parents' presence to assist with continuity of care and find them to be a source of comfort.
Transitioning leads to evolving youth's role	The role of the adolescent and young adult transforms as they prepare or transition to adult healthcare.
Identifying barriers	Adolescents and young adults have recognized differences between the pediatric and adult setting, which causes concern for them during the transition.
Lacking transition readiness	A lack of transition preparation can lead to a variety of poor outcomes for youth in terms of health outcomes, relationships with new providers and their parents, and their perspective of the transfer to the adult healthcare setting.
Recommendations for improvement to transition preparation	Adolescents and young adults have numerous suggestions for assisting them as they plan for transition, which they would like to be implemented prior to transition to the adult healthcare setting.

Table 6 *Deficiencies and Recommendations Reported by AYA*

Six Core Elements of Health Care Transition™	Deficiencies Reported by AYA	Recommendations Reported by AYA Ready for Practice Translation
Transition Policy	<ul style="list-style-type: none"> • Not told far enough in advance about transition • Report unsystematic transfers • Do not know what to expect with transition 	<ul style="list-style-type: none"> • Provide details about process and resources during transition • Talk about transition beginning at early ages • Provide a structured program to encourage preparation
Transition Tracking and Monitoring	<ul style="list-style-type: none"> • Not discussed by AYA in primary studies 	<ul style="list-style-type: none"> • Not discussed by AYA in the primary studies
Transition Readiness	<ul style="list-style-type: none"> • Lack knowledge and ability to describe their disease condition, history, and current treatments • Not ready for disease self-management • Do not know when to seek care • Do not feel emotionally ready • Caregivers have trouble letting go of responsibilities 	<ul style="list-style-type: none"> • Provide repetitive education about disease conditions to help AYA become more knowledgeable about their medical conditions and needs • Encourage and support AYA to gradually assume more independence and responsibility as they grow up • Base transition upon individual readiness and not just age • Make AYA active participants in their pending transfer and encourage caregivers to support new roles for AYA
Transition Planning	<ul style="list-style-type: none"> • Do not know who are their healthcare resources post-transition • AYA reported not knowing how to navigate insurance or how to obtain appointments with their insurance 	<ul style="list-style-type: none"> • Encourage AYA to try out providers • Encourage active, independent AYA role in clinic appointments earlier • Utilize peers to help provide insight into what to expect • Provide age pertinent information and resources (i.e. information on drugs, alcohol, pregnancy, lifestyle) • Provide information on transition in a creative method (i.e. workshop, shadowing, programming)
Transfer of Care	<ul style="list-style-type: none"> • Do not know how to identify new providers • Report poor communication between pediatric and adult providers 	<ul style="list-style-type: none"> • Provide assistance in identifying providers • Coordinate care between pediatric and adult providers or offer “joint clinics”
Transfer Complete	<ul style="list-style-type: none"> • Found adult providers’ expectations were unreasonable • Report having poor experiences with new adult providers and subsequently developing mistrust of the new providers • Reported periods of interruption to care • Reported negative feelings regarding environment or approach with first visits to adult providers settings 	<ul style="list-style-type: none"> • Encourage adult practices to tailor their care towards AYA and their needs • Develop collaborations between pediatric and adult practices to ensure continuity of care • Work with adult providers to reduce inconsistencies in care post-transition • Encourage adult provider to have person-centered communication that is appropriate for youth adults • Maintain pediatric provider role after transition to ensure success

Supplement 1

Characteristics Across Studies

Author	Chronic Disease Type	Study Aim	Method	n	Context*	Summary of Findings
Allen, Channon, Lowes, Atwell, & Lane (2011)	Diabetes mellitus	“To explore the experiences of young people and their carers during the transition from child to adult diabetes services”	Thematic analysis	46	United Kingdom Recruited from clinics 23 F/ 23 M 13-22 years Individual interviews	There are two separate transitions coexisting: “transition from pediatric to adult care” and “the child’s move from dependence on caregivers to independence.”
Asp, Bratt, & Bramhagen (2015)	Congenital heart disease	“The aim of the study was to explore the experiences of young adults with congenital heart disease in Sweden after transfer from the pediatric cardiac clinic to the adult congenital heart disease clinic.”	Content analysis	16	Sweden Recruited from clinics Pain < 6 months 11 F/ 5 M 20-24 years Individual interviews	The analysis identified five themes: “feeling secure during the transfer process,” “experiencing trust in the care, expecting to be involved,” “assuming responsibility for one’s health is a process,” and “lack of knowledge leads to uncertainty.”
Bemrich-Stolz, Halanych, Howard, Hilliard, & Lebensburger (2015)	Sickle cell disease	“The purpose of this qualitative research study was to identify barriers to transition to adult care by exploring the transition	Content analysis	10	United States Recruited a clinic 7 F/ 3 M 24-55 years Individual interviews	Participants reporting being unprepared for the transition. Participants reported feelings of physician mistrust, challenges with new employers, insurance difficulties, and personal relationship struggles. Transition

Author	Chronic Disease Type	Study Aim	Method	n	Context*	Summary of Findings
Bomba, Herrmann-Garitz, Schmidt, Schmidt, & Thyen (2017)	Diabetes mellitus, cystic fibrosis, inflammatory bowel disease	experiences from the perspective of adult patients with sickle cell disease.” “To investigate the experiences and needs in the transition phase of adolescents with chronic conditions.”	Content analysis	29	Germany and Switzerland Recruited from clinics 17 F/ 12 M 15-27 years Individual interviews	did bring positive experiences including feelings of increased self-efficacy, autonomy, and self-care. Themes identified included: “transfer to adult medicine,” “their new role as a patient,” “orientation within the healthcare system,” “vocational issues,” “detachment from parents,” “social support,” “contraception,” “substance abuse,” “family planning stress-management,” “activation of resources,” and “developing personal goals.”
Burstrom, Ojmy-Joelsson, Bratt, Lundell, & Nisell (2016)	Congenital heart disease	“To identify and describe the needs of adolescents with CHD and their parents during the transition before transfer to adult cardiologic healthcare.”	Content analysis	13	Sweden Recruited via advertisement 7 F/6 M 16-18 years Individual interviews	Themes identified included: “change of relationships,” “knowledge and information,” and “daily living.”
Bustrom, Bratt, Frenckner, Nisell, Hanseus,	Congenital heart disease	“To explore what adolescents with congenital heart disease view as important in the	Content analysis	17	Sweden Recruited from university hospitals	The analysis revealed one main category (becoming a manager of the condition) and four subcategories (sufficient knowledge about the health, be a

Author	Chronic Disease Type	Study Aim	Method	n	Context*	Summary of Findings
Rydberg, & Ojmyr-Joelsson (2017)	Cerebral palsy	preparation for the transfer to adult care."	Phenomenological analysis	9	10 F/7 M 14-18 years Focus groups	participant in the care, parental support, and communicate with others about health).
Carroll 2015)		"To uncover the meaning of transition to adult-centered care as experienced by young adults with cerebral palsy."			United States Recruited using an advertisement 6 F/3 M 19-25 years Individual interviews	Five main themes were identified: "expert novice," "evidence and experience-based expectations," "negotiating new systems," "interdependence," and "accepting less."
Catena, Rempel, Kovacs, Rankin, Muhll, & Mackie (2018)	Congenital heart disease	"To understand the perspectives of young adult patients who have been transferred to adult healthcare versus those who continue to received pediatric care."	Content and thematic analysis	21	Canada Recruited from clinics 9 F/12 M 18-25 years Individual interviews	Participants reported a difference between pediatric and adult settings. Participants felt there would be an adjustment period after transition. Transition did bring positive aspects such as increasing responsibility and independence.
DiFazio, Harris, Vessey, Glader, & Shanske (2014)	Cerebral palsy	"To describe and define the experience of adults with cerebral palsy and parents who have bene involved in a transfer of physiatry care from pediatric to adult healthcare and to explore their	Content analysis	5	United States Recruited from an academic hospital 3 F/2 M 22-44 years Focus groups	Themes identified in the study included: "lost in transition," "roadmap to care," "list of none," and "one stop shopping." Participants felt they were left behind during transition, wanted a plan prior to transition, did not feel they could access adult providers, and needed recommendations for transition.

Author	Chronic Disease Type	Study Aim	Method	n	Context*	Summary of Findings
Dupuis, Duhamel, & Gendron (2011)	Cystic fibrosis	experiences more generally in the transition." "To explore the experiences of adolescents with cystic fibrosis and their parents prior to transfer to adult healthcare."	Content analysis	7	Canada Recruited from a clinic 5 F/2 M 15-18 years Individual interviews	Themes identified in the study included: "parents and adolescents living with CF; a dynamic of normalization," "parents confronted with adolescent's urgent desire to live," "parents experienced ongoing suffering," and "suffering unrevealed to health care provider."
Fair, Goldstein, & Dizney (2015)	HIV	"To examine the congruence of transition expectations of adolescents living with HIV and their guardians."	Grounded theory	18	United States Recruited from a clinic 11 F/7 M 15-19 years Individual interviews	Participants and their caregivers disagreed about upon what should happen during transition. They also disagreed about caregiver level of involvement. They wanted providers to address these disagreements.
Freeman, Stewart, Cunningham, & Gorter (2018)	Cerebral palsy	"To ask adults with cerebral palsy between the ages of 20 and 40 to reflect on information they received during the transition to adulthood."	Content analysis	9	Canada Recruited through advertisements 7 F/2 M 25-38 years Individual interviews	Themes identified in the study included: "recognizing and supporting information needs," "getting creative (strategies young people use when confronted with environmental barriers when seeking information)," and "gaps and advice for the future."
Garvey, Beste, Luff, Atakov-	Diabetes mellitus	"To explore the experiences of	Thematic analysis	26	United States	Themes identified in the study included: "non-purposeful

Author	Chronic Disease Type	Study Aim	Method	n	Context*	Summary of Findings
Castillo, Wolpert, & Ritholz (2014)		transition from pediatric to adult diabetes care reported by post-transition emerging adults with diabetes."			Recruited from a clinic 16 F/10 M Mean age 20.3±3.2 years Focus groups	transition (patients reported a lack of transition preparation by providers)," "vulnerability in the college years (patients reported a lack of follow-up, risky behaviors, and management problems)," "unexpected differences between the settings," and "patients' wish list for improving the transition process."
Hilderson, Eyckmans, Von der Elst, Westhovens, Wouters & Moons (2012)	Juvenile idiopathic arthritis	"To explore the experiences and expectations of transferred young adults with JIA, regarding their transfer from pediatric to adult care."	Content analysis	11	Belgium Rheumatology clinic 8 F/3 M 20-30 years Individual interviews	Themes identified in the study included: "preparation," "parental involvement," and "an adapted setting for the late-adolescent or early adult."
Hilliard, Perlus, Clark, Haynie, Plotnick, Guttman-Bauman, & Iannotti (2014)	Diabetes mellitus	"To understand the concerns, expectations, preferences, and experiences of pre-transition adolescents and parents and post-transition young adults."	Thematic coding	60	United States Recruited from clinics Unknown 15-22 years Individual interviews	Themes identified in the study included: "timing of transfer," "early preparation for transition," "the desire for developmentally appropriate interactions with providers," "the maintenance of family and social support," and "strategies for coordinating care between

Author	Chronic Disease Type	Study Aim	Method	n	Context*	Summary of Findings
Huang, Gottschalk, Pian, Dillon, Barajas, & Batholomew (2011)	Inflammatory bowel disease, cystic fibrosis, and diabetes mellitus	“To examine the process of transition from pediatric to adult healthcare services from the perspective of young adults with chronic disease.”	Content and narrative analysis	10	United States Recruited from clinics 6 F/ 4 M 18-25 years Focus group	pediatric and adult care providers.” Themes identified in the study included: “transition experiences in the context of relationship between patients, parents, and healthcare providers,” “differences between pediatric and adult-oriented medicine and how these differences inhibit or facilitate transition,” and “identifying transition services that should be provided to young patients with chronic disease.”
Lewis & Noyes (2013)	Epilepsy	“To explore the views of young people with epilepsy about their experiences of communication, information, and knowledge exchange in two epilepsy services during transition.”	Thematic analysis	30	United Kingdom Recruited from clinics Not specified 13-19 years Individual interviews, small group interviews, and focus groups	A coordinated, joint epilepsy clinic helped to facilitate youth’s engagement with their providers. Things that were important to successful transition included continuity of care, ongoing support, and age-appropriate communication. Parents did not have the skills or knowledge necessary to support independence in their child.
Lochridge, Wolff, Oliva, & O’Sullivan-	Solid organ transplant	“To understand the perceptions of patients and their parents about self-	Phenomenological	10	United States Recruited from an	Themes identified in the study included: “negative perceptions of adult transplant centers due to anxiety regarding the

Author	Chronic Disease Type	Study Aim	Method	n	Context*	Summary of Findings
Oliveira (2013)		care management during transition."			academic hospital 3 F/7 M 16-22 years Individual interviews	competencies of new providers in caring for the child's medical needs," "unfamiliarity with adult medical providers," "fear of letting go and assuming greater responsibility," and "discrepancies between providers, parents, and patients."
Lopez, Karlsten, Bonaduce, Nigris, King, Saliccioli, Jiang, Marelli, Kovacs, Fordis, & Thompson (2015)	Congenital heart disease	"To explore the transition process in congenital heart disease care through the perceived needs and concerns of adolescents and young adults."	Content analysis	20	United States Recruited via advertisement 9 F/11 M 16-20 years Individual interviews	Majority of adolescents wanted more disease information and were concerned with becoming an adult. Adolescents and young adults were interested in peer mentorship. They wanted more information on long-term complications of CHD, employment, insurance, family planning, and mental health.
Morsa, Lombrail, Boudailliez, Godot, Jeantils, & Gagnayre (2018)	Multiple chronic diseases	"To identify education needs related to the development of self-care skills, informational needs related to managing health behaviors, and psychosocial needs relating to young people's coping	Thematic analysis	17	France Recruited from hospitals 10 F/7 M Mean age 18.2 years Individual interviews	Themes identified in the study included: "learning how to have a new role," "learning how to adopt a new lifestyle," "learning how to use a new health care service," "maintaining a dual relationship with pediatric and adult care," and "having experience sharing with peers."

Author	Chronic Disease Type	Study Aim	Method	n	Context*	Summary of Findings
		mechanisms.”				
Nicholas, Kaufman, Pinsk, Samuel, Hamiwka, & Molzahn (2018)	Chronic kidney disease	“To explore the perceptions and experiences of young people with CKD and their families regarding transition.”	Content analysis	28	Canada Recruited from clinics 12 F/ 16 M 12-25 years Individual interviews	Themes identified in the study included: “parental challenge relinquishing care responsibility,” “synergistic and divergent expectations between youth and parents,” “tensions in youth self-care readiness,” “desired healthcare provider roles in transition preparation,” and “system considerations in transition.”
Nguyen, Henderson, Stewart, Hlyva, Punthakee, & Gorter (2016)	Multiple chronic diseases	“To explore self-management from the perspectives of young, parents, and healthcare providers in transition to adult healthcare.”	Interpretive phenomenological approach	11	Canada Recruited from clinics 6 F/5 M 18-21 years Individual interviews	Themes identified in the study included: “increasing independence of youth,” “parents as safety nets,” and “healthcare providers as enablers and collaborators.”
Plevinsky, Gumidyala, & Fishman (2014)	Inflammatory bowel disease	“To understand transition of young adults with IBD, the impact of the pediatric patient-provider relationships, and what determines the adult patient-	Phenomenology approach	29	United States Recruited with advertisements Unknown 18-30 years Qualitative questionnaires	There were positive themes revolving around autonomy, independence, and trust. There were negative themes associated with discomfort with initial visits and challenges with logistics.

Author	Chronic Disease Type	Study Aim	Method	n	Context*	Summary of Findings
Porter, Graff, Lopez, & Hankins (2014)	Sickle cell disease	provider relationship.” “To describe the perspectives of adolescents with SCD, siblings, and their caregivers regarding the transition from pediatric to adult SCD care to identify recommendations for improving the transition process.”	Content analysis	11	United States Recruited with letters Genders unknown 12-18 years Focus groups	Adolescents expressed awareness that they would need to take on increased responsibility. They reported experiencing many emotions regarding transition including: fear, sadness, and worried. They expressed reluctance to leave the pediatric setting and expressed doubt in the adult providers abilities. They recommended pediatric providers help them practice independence, increase familiarity with the adult providers and allow them to meet with peers living with SCD.
Porter, Wesley, Zhao, Rupff, & Hankins (2017)	Sickle cell disease	“To explore perspectives of transition and transition readiness of young adults with SCD who have transitioned to adult health care.”	Content analysis	19	United States Recruited from 12 F/ 7 M 18-30 years Focus groups	Themes were consistent with the SMART model. There was a focus on adult provider relationships and negative medical experiences. The most important issues which young adults wanted information on included: “identifying an adult provider,” “seeking emergency care,” “understanding SCD treatment,” “knowing SCD complications,” and “being

Author	Chronic Disease Type	Study Aim	Method	n	Context*	Summary of Findings
Price, Corbett, Lewis-Barned, Oliver, & Dovey-Pearce (2011)	Diabetes mellitus	“To evaluate a “transitions pathway services” in diabetes and to explore elements which young people themselves regard as important.”	Thematic analysis	11	United Kingdom Recruited from clinic Genders unknown 16-18 years Individual interviews	aware of the effect of health behaviors”. Themes identified in the study included: “the transition process,” “the experience and organization of transfer,” “organization of services,” “information and education,” “the healthcare consultation,” and “the need to be inclusive of young people’s needs.”
Ridosh, Braun, Roux, Bellin, & Sawin (2011)	Spina bifida	“To examine the transition to adulthood in young adults with spina bifida and to explore condition-related needs and life skills required during the transition process.”	Thematic analysis	10	United States Recruited from clinics 4F / 6M 18-25 years Individual interviews	Themes identified in the study included: “struggling for independence,” “limiting social interactions and experiences with stigma,” and “building inner strength.”
Ritholz, Wolpert, Beste, Luff, & Garvey (2014)	Diabetes mellitus	“To explore perceptions that emerging adults with type 1 diabetes have of their patient-provider relationships across the transition from pediatric to adult care.”	Thematic analysis	26	United States Recruited from clinics 16 F/ 10M Mean ages 26.2±2.5 years Focus groups	Themes identified in the study included: “loss and gain in provider relationships across the transition,” “partners in care versus one’s own – patients’ valued how adult providers’ collaborative conversations promoted their involvement and accountability,” and “improving provider approaches to transition

Author	Chronic Disease Type	Study Aim	Method	n	Context*	Summary of Findings
Sobota, Umeh, & Mack (2015)	Sickle cell disease	"To learn from young adults with sickle cell disease about their experiences leaving pediatric care and perspectives on what makes a successful transition."	Grounded theory	15	United States Recruited from clinics 8 F / 7 M 18-28 years Focus groups	– patients recommended that pediatric providers actively promote emerging adults' autonomy while maintaining parental support, communication with adult providers, and follow-up with transitioning patients." Themes identified in the study included: "facilitators of transition," "barriers to transition," "what young adults wished for in a transition program," and "how they define a successful transition."
Tierney, Deaton, Jones, Oxley, Biesty, & Kirk (2013)	Cystic fibrosis	"To explore young people's experiences of transferring."	Thematic analysis	19	United Kingdom Recruited from clinics 7 F/12 M 17-19 years Individual interviews	Transfer was a period of flux where the participants moved from a prescriptive service to one that called for autonomy. Rites of passage, such as being visited by a member of the adult team and a few appointment within this new setting. Without rites of passage progression can be hindered towards becoming an adult patient.
Wright, Elwell, McDonagh,	Liver transplant	"To explore the views and perceptions of young people with liver	Interpretive phenomenological analysis	17	United Kingdom Recruited from clinics	Two main themes emerged: "relationships with adult healthcare providers" and "continuity of care." Under the

Author	Chronic Disease Type	Study Aim	Method	n	Context*	Summary of Findings
Kelly, & Wray (2016)		transplants who were about to, or who had already, transferred to adult care to understand their experiences of transition and to elicit what they believe constitutes a “good” transition.”			10 F/7 M 15-25 years Individual interviews	theme relationships with adult healthcare providers included: “they don’t know me” and “it was sad to go.” Under the theme continuity of care there were two subthemes: “fear of the unknown” and “it’s like meet and greet again.”

*Context

- Country in which study undertaken
- Process of recruitment
- Gender of participants
- Age range
- Process of data collection

Supplement 2

CASP Appraisal of Included Studies

Author and Citation	Clear Statement of the Aims	Qualitative Methodology Appropriate	Research Design Appropriate	Recruitment Strategy Appropriate	Data Collection Addressed	Relationships Adequately Considered	Ethical Issues were Considered	Data Analysis Sufficiently Rigorous	Clear Statement of Findings	Research is Valuable
Allen et al., (2011)	Yes	Yes	Can't Tell	Yes	Can't Tell	Can't Tell	Can't Tell	Yes	Yes	Yes
Asp et al., (2015)	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes
Bemrich-Stolz et al., (2015)	Yes	Yes	Yes	Yes	Yes	Can't Tell	Yes	Yes	Yes	Yes
Bagatell et al., (2017)	Yes	Yes	Yes	Can't Tell	Yes	Can't Tell	Can't Tell	Yes	Yes	Yes
Bomba et al., (2017)	Yes	Yes	Yes	Can't Tell	Can't Tell	Can't Tell	Yes	Can't Tell	Yes	Can't Tell
Bustrom et al., (2016)	Yes	Yes	Yes	Yes	Yes	No	Yes	Yes	Yes	Yes
Bustrom et al., (2017)	Yes	Yes	Yes	Yes	Yes	No	No	Can't Tell	Yes	Yes
Carroll (2015)	Yes	Yes	Yes	Yes	Yes	Can't Tell	No	Can't Tell	Yes	Can't Tell
Catena et al., (2018)	Yes	Yes	Yes	Yes	Yes	Can't Tell	Yes	Yes	Yes	Yes
DiFazio et al., (2014)	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes
Dupuis et al., (2011)	Can't Tell	Yes	Yes	Yes	Yes	No	Yes	Yes	Yes	Yes
Fair et al., (2015)	Yes	Can't Tell	Yes	Yes	Can't Tell	Can't Tell	Yes	Can't Tell	Yes	Can't Tell
Freeman et al., (2018)	Yes	Yes	Yes	Can't Tell	Can't Tell	Can't Tell	Yes	Yes	Yes	Yes
Garvey et al., (2014)	Yes	Yes	Yes	Yes	Yes	Can't Tell	Yes	Yes	Yes	Yes

Hilderson et al., (2012)	Yes	Can't Tell	Yes	Yes	Yes	Can't Tell	Yes	Yes	Yes	Yes
Hilliard et al., (2014)	Yes	Yes	Yes	Yes	Yes	No	Yes	Can't Tell	Yes	Yes
Huang et al., (2011)	Yes	Yes	Yes	Yes	Yes	Can't Tell	Yes	Yes	Yes	Yes
Lewis & O2 (2013)	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes
Lochridge et al., (2013)	Yes	Yes	Can't Tell	Yes	Can't Tell	No	No	Can't Tell	Yes	Can't Tell
Lopez et al., (2015)	Yes	Yes	Yes	Yes	Can't Tell	No	Yes	Can't Tell	Can't Tell	Can't Tell
Morsa et al., (2018)	No	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes
Nicholas et al., (2018)	Yes	Yes	Yes	Can't Tell	Can't Tell	Yes	Yes	Yes	Yes	Yes
Nguyen et al., (2016)	Yes	Yes	Yes	Yes	Yes	Can't Tell	Can't Tell	Yes	Yes	Yes
Plevinsky et al., (2014)	Yes	Yes	Yes	Yes	Can't Tell	Can't Tell	Yes	Can't Tell	Can't Tell	Can't Tell
Porter et al., (2014)	Yes	Yes	Yes	Yes	Yes	Can't Tell	Yes	Yes	Yes	Yes
Porter et al., (2017)	Yes	Yes	Can't Tell	<u>Yes</u>	Can't Tell	Can't Tell	Can't Tell	Yes	Yes	Yes
Price et al., (2011)	Yes	Yes	Yes	Yes	Yes	Can't Tell	No	Yes	Yes	Yes
Ridosh et al., (2011)	Yes	Yes	Yes	Yes	Yes	Can't Tell	Yes	Yes	Yes	Yes
Ritholz et al., (2014)	Yes	Yes	Can't Tell	Yes	Yes	Can't Tell	Yes	Yes	Yes	Yes
Sobota et al., (2015)	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes
Tierney et al., (2013)	Yes	Yes	Yes	Yes	Yes	Can't Tell	Yes	Yes	Yes	Yes
Wright et al. (2016)	Yes	Yes	Yes	Yes	Can't Tell	Can't Tell	Can't Tell	Can't Tell	Yes	Yes

Supplement 3

CERQual Qualitative Evidence Profile

Meta-theme	Studies contributing	Methodological limitations	Relevance	Coherence	Adequacy	Overall	Explanation
Developing transition readiness	Allen et al., (2011) Asp et al., (2015) Benrich-Stolz et al., (2015) Bomba et al., (2017) Burström et al., (2016) Burström et al., (2017) Catena et al., (2018) Hilderson et al., (2013) Hilliard et al., (2014) Lewis & Noyes, (2013) Lochridge et al., (2013) Lopez et al., (2015) Morsa et al., (2018) Nicholas et al., (2018) Porter et al., (2014) Porter et al., (2017) Ritholz et al., (2014) Sobota et al., (2015)	Minor concerns about methodological limitations	Minor concerns about relevance UK, Sweden, US, Germany, Switzerland, Belgium, Canada, France 162/298 Outpatient clinics, Advertisements, Tertiary care (8 countries represented in the studies; 54% participants were female)	Minor concerns about coherence DM, CHD, SCD, CF, IBD, MS, HIV, CKD, Juvenile Idiopathic Arthritis, Epilepsy, solid organ transplant (Theme consistent across 18 studies and 11 chronic conditions)	Minor concerns about adequacy (Themes richly described across 14/18 studies)	High confidence	This finding was graded as high confidence with studies having minor methodological limitations. The findings are only slightly more relevant to females but relevant across eight cultures and across a range of contexts. Coherence across chronic conditions was high and themes were richly described across most studies.

Conceiving expectations based upon pediatric healthcare	<p>Asp et al., (2015)</p> <p>Burström et al., (2016)</p> <p>Carroll et al., (2015)</p> <p>Catena et al., (2018)</p> <p>DiFazio et al., (2014)</p> <p>Garvey et al., (2014)</p> <p>Hilliard et al., (2014)</p> <p>Huang et al., (2011)</p> <p>Lewis & Noyes, 2013)</p> <p>Lochridge et al., (2013)</p> <p>Morsa et al., (2018)</p> <p>Plevinsky et al., (2015)</p> <p>Porter et al., (2014)</p> <p>Porter et al., (2017)</p> <p>Price et al., (2011)</p> <p>Ritholz et al., (2014)</p> <p>Tierney et al., (2013)</p> <p>Wright et al., (2016)</p>	Minor methodological limitations	<p>Moderate concerns about relevance</p> <p>Sweden, US, UK, France</p> <p>Outpatient care, advertisement, tertiary care</p> <p>116/219</p> <p>(4 countries represented in the studies; 53% female)</p>	<p>Minor concerns about coherence</p> <p>CHD, DM, IBD, CF, MS, Epilepsy, solid organ transplant, HIV, SCD, liver transplant, Cerebral Palsy</p> <p>(Consistent across 18 studies and 11 chronic conditions)</p>	<p>Moderate concerns about adequacy</p> <p>(Themes richly described across 14/18 studies)</p>	Moderate confidence	<p>This finding was graded as moderate confidence in relation to relevance and adequacy and high confidence in relation to methodological limitations and coherence. The findings are only slightly more relevant to females and across a range of contexts but only relevant across four cultures. Coherence across chronic conditions was high, but themes were only richly described across a moderate number of the studies.</p>
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Transitioning leads to evolving parent role	<p>Allen et al., (2011) Asp et al., (2015) Burstrom et al., (2016) Burstrom et al., (2017) Carroll et al., (2015) Catena et al., (2018) Hilderson et al., (2013) Hilliard et al., (2014) Lewis & Noyes, (2013) Lochridge et al., (2013) Morsa et al., (2018) Nicholas et al., (2018) Plevinsky et al., (2015) Porter et al., (2017) Ridosh et al., (2011) Sobota et al., (2015) Tierney et al., (2013) Wright et al., (2016)</p>	Minor methodological limitations	<p>Moderate concerns about relevance</p> <p>UK, Sweden, Belgium, US, Canada, France</p> <p>Outpatient clinic, advertisement, tertiary care</p> <p>140/268</p> <p>(6 countries represented; 52% female)</p>	<p>Minor concerns about coherence</p> <p>DM, CHD, HIV, MS, CKD, Juvenile Idiopathic Arthritis, Epilepsy, solid organ transplant, IBD, Spina Bifida, SCD, CF, liver transplant, Cerebral Palsy</p> <p>(Consistent across 18 studies and 14 chronic conditions)</p>	<p>Minor concerns about adequacy</p> <p>(Themes well described across 13/18 studies)</p>	High confidence	<p>This finding was graded as high confidence because there were minor methodological limitations, moderate concerns about relevance. The findings were fairly even relevance across genders and across a range of contexts. The findings were coherent across 14 chronic conditions and themes were adequately described across a large number of the studies.</p>
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Transitioning leads to evolving youth role	Asp et al., (2015) Catena et al., (2018) DiFazio et al., (2014) Hildersen et al., (2015) Huang et al., (2011) Lewis & Noyes (2013) Lochridge et al., (2013) Lopez et al., (2015) Morsa et al., (2018) Nicholas et al., (2018) Nguyen et al., (2016) Plevinsky et al., (2015) Porter et al., (2014) Porter et al., (2017) Price et al., (2011) Ridosh et al., (2011) Ritholz et al., (2014) Sobota et al., (2015) Tierney et al., (2013)	Minor methodological limitations	Moderate concerns about relevance Sweden, Belgium, US, Canada, UK, France Outpatient clinic, advertisement, tertiary care 124/238 (6 countries represented; 52% female)	Minor concerns about coherence CHD, Juvenile Idiopathic Arthritis, DM, solid organ transplant, IBD, SCD, Spina Bifida, CF, Epilepsy, Cerebral Palsy, MS, HIV, CKD (Consistent across 19 studies and 13 chronic conditions)	Moderate concerns about adequacy (Themes richly described across 13/19 studies)	Moderate confidence	This finding was graded as moderate confidence in relation to relevance and adequacy and graded as high in relation to coherence and methodological limitations. The finding was fairly even across genders and across a range of contexts. The finding was coherent across 13 chronic conditions and the themes were richly described across a moderate number of the studies.
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Identifying barriers	<p>Bemrich-Stolz et al., (2015)</p> <p>Hilliard et al., (2014)</p> <p>Lochridge et al., (2013)</p> <p>Porter et al., (2014)</p> <p>Price et al., (2011)</p> <p>Tierney et al., (2013)</p> <p>Wright et al., (2016)</p> <p>(Catena et al., 2018)</p> <p>(Porter et al., 2017)</p> <p>(Carroll et al., 2015)</p> <p>(DiFazio et al., 2014)</p>	Minor methodological limitations	<p>Moderate concerns about relevance</p> <p>US, UK, Canada</p> <p>Outpatient clinic, advertisement, tertiary care</p> <p>57/110</p> <p>(3 countries represented; 52% female)</p>	<p>Moderate concerns about coherence</p> <p>SCD, DM, solid organ transplant, CF, liver transplant, CHD, Cerebral Palsy</p> <p>(Consistent across 11 studies and 7 chronic conditions)</p>	<p>Minor concerns about adequacy</p> <p>(Themes richly described across 8/11 studies.)</p>	Moderate confidence	<p>This finding was graded as moderate confidence in relation to the relevance, and coherence. It was graded high confidence in relation to the methodological limitations and adequacy. The findings were fairly even across gender and a range of contexts. There was moderate coherence as the findings were only described across 7 conditions; however, they were richly described across a majority of the studies which discussed the theme.</p>
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Lacking transition readiness	<p>Asp et al., (2015) Bemrich-Stolz et al., (2015) Bomba et al., (2017) Burstrom et al., (2016) Burstrom et al., (2017) Carroll et al., (2015) Catena et al., (2018) DiFazio et al., (2014) Garvey et al., (2014) Hilderson et al., (2013) Huang et al., (2011) Lewis & Noyes, (2013) Lochridge et al., (2013) Lopez et al., (2015) Morsa et al., (2018) Plevinsky et al., (2015) Porter et al., (2017) Price et al., (2011) Ridosh et al., (2011) Sobota et al., (2015) Tierney et al., (2013) Wright et al., (2016)</p>	Minor methodological limitations	<p>Minor concerns about relevance</p> <p>Sweden, US, Germany, Switzerland, Belgium, UK, Canada, France</p> <p>Outpatient care, advertisement, tertiary care</p> <p>175/310</p> <p>(8 countries represented; 56% female)</p>	<p>Minor concerns about coherence</p> <p>CHD, SCD, DM, Juvenile Idiopathic Arthritis, IBD, CF, Epilepsy, solid organ transplant, HIV, MS, liver transplant, Cerebral Palsy</p> <p>(Consistent across 22 studies and 12 chronic conditions)</p>	<p>Minor concerns about adequacy</p> <p>(Themes richly described across 19/22 studies.)</p>	High confidence	<p>This finding was graded as high confidence with studies having minor methodological limitations. The findings are only slightly more relevant to females but relevant across eight cultures and across a range of contexts. Coherence across chronic conditions was high and themes were richly described across most studies.</p>
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Recommendations for improvements	<p>Bemrich-Stolz et al., (2015)</p> <p>Bomba et al., (2017)</p> <p>Burström et al., (2016)</p> <p>Burström et al., (2017)</p> <p>Carroll et al., (2015)</p> <p>Catena et al., (2018)</p> <p>DiFazio et al., (2014)</p> <p>Freeman et al., (2018)</p> <p>Garvey et al., (2014)</p> <p>Hilderson et al., (2013)</p> <p>Hilliard et al., (2014)</p> <p>Huang et al., (2011)</p> <p>Lewis & Noyes (2013)</p> <p>LochrIDGE et al., (2013)</p> <p>Lopez et al., (2015)</p> <p>Morsa et al., (2018)</p> <p>Nicholas et al., (2018)</p> <p>Nguyen et al., (2016)</p> <p>Porter et al., (2017)</p> <p>Price et al., (2011)</p> <p>Wright et al., (2016)</p>	Minor methodological limitations	<p>Minor concerns about relevance</p> <p>US, Germany, Switzerland, Sweden, Belgium, UK, Canada, France</p> <p>Outpatient clinic, advertisement, tertiary care</p> <p>158/282</p> <p>(7 countries represented; 56% female)</p>	<p>Minor concerns about coherence</p> <p>SCD, DM, CF, IBD, CHD, DM, Juvenile Idiopathic Arthritis, Epilepsy, solid organ transplant, HIV, liver transplant, Cerebral Palsy, MS, HIV, CKD</p> <p>(Consistent across 21 studies and 15 chronic conditions)</p>	<p>Moderate concerns about adequacy</p> <p>(Themes richly described across 16/21 studies.)</p>	High confidence	<p>This finding was graded as high confidence in relation to methodological limitations, relevance, and adequacy and moderate confidence in coherence. There were minor methodological limitations. It was slightly more relevant to females. There was high coherence across 15 chronic conditions and the themes were richly described in a moderate number of the studies.</p>
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CHAPTER FOUR

METHOD

The following research proposal is a modified version of the proposal which was submitted for and successfully funded by the National Institute of Nursing Research Ruth L Kirchstein Predoctoral Individual National Research Service Award for 2019-2020.

Specific Aims

Sickle cell disease (SCD) is a debilitating inherited disease which impacts about 100,000 individuals in the United States (US). The incidence of SCD is 15.5 per 1,000 newborns (Ojodu et al., 2014). A majority of those affected are African-American and living in poverty, which impacts their access to care (National Heart, 2018; Raphael et al., 2009). Costs to the US health system are estimated to be \$488 million annually (National Heart, 2018). Young adults with SCD face higher rates of morbidity and mortality in the period shortly after transitioning to the adult healthcare setting due to inadequate preparation for the change to adult healthcare setting (Blinder et al., 2013; Dickerson, Klima, Rhodes, & O'Brien, 2012; Quinn, Rogers, McCavit, & Buchanan, 2010). Currently, over 93% of children with SCD will live into adulthood and require transition to the adult healthcare setting, creating a need to find solutions to effectively prepare young adults with SCD for this transition (Quinn et al., 2010).

Successful transition has become a priority in the area because the SCD comorbidities increases with age, in particular acute care utilization sharply increases after 18, and there is an alarming increase in mortality for young adults (Blinder et al., 2013; Brousseau, Owens, Mosso, Panepinto, & Steiner, 2010; Lanzkron, Carroll, & Haywood, 2013; Paulukonis et al., 2014; Quinn et al., 2010). Young adults with SCD just

after transition have the highest rates of frequent acute care visits (> 10 acute care visits per year) due to SCD complications (Dickerson et al., 2012; Quinn et al., 2010). After transition to adult healthcare settings there is an inappropriate reduction in the use of medical treatment. Young adults receive fewer recommended transfusions, which increases their risk of neurological complications, acute chest syndrome, and anemia (Blinder et al., 2013). Fewer hydroxyurea prescriptions are filled, which increases their risk for pain crises (Blinder et al., 2013). Most concerning is the increase in mortality which occurs after transition with a longitudinal study showing that in a study of 940 youth with SCD, 85% of the deaths occurred after transition with a mean time to death being 1.8 years after transition (Blinder et al., 2013). Transition preparation has received increased attention as a protection against adverse outcomes as adequate transition preparation minimizes disruptions in treatment. There is a critical need for a structured process which focuses on preparing adolescents and young adults (AYA) with SCD based on psychological development and achievement of milestones related to disease-specific knowledge and self-management skills (DeBaun & Telfair, 2012; Dickerson et al., 2012; Edwards, Telfair, Cecil, & Lenoci, 2000; Elander, Lusher, Bevan, & Telfer, 2003; Lisa A. Schwartz et al., 2014; L. A. Schwartz, Tuchman, Hobbie, & Ginsberg, 2011).

The main maturational milestone experienced by AYA is that of gaining independence from their families, but this milestone is impacted by numerous developmental, environmental, and socioeconomic challenges (Anderson et al., 2009; Hanna, Juarez, Lenss, & Guthrie, 2003; Miller & Drotar, 2006; Pai et al., 2010; J. L. Ryan, Arnett, Pai, & Modi, 2014). Anchored by The Individual and Family Self-

Management Theory, this study will assess AYA's decision-making involvement (ways in which AYAs contribute to the decision-making process regarding disease management) and its relationship to AYA's self-efficacy (level of confidence in managing their SCD) and transition readiness (disease-specific knowledge and self-management skills necessary for successful transition) (Edwards et al., 2000; Gilleland, Amaral, Mee, & Blount, 2012; Miller & Drotar, 2006). While self-efficacy in SCD self-management has been found to be positively related to transition readiness in AYAs it is less clear that improved self-efficacy alone changes AYA's capability to manage their disease (Treadwell et al., 2016). AYA decision-making involvement has not been studied for its relationship to transition readiness but could potentially be a modifiable factor that would influence transition readiness. Prior work in AYA decision-making involvement suggests it can enhance self-efficacy in individual and disease self-management (Liprie, 1993; Miller & Drotar, 2006; P. H. White, McManus, McAllister, & Cooley, 2012). AYAs do not make decisions isolated from family and there is the potential that AYAs' growing independence from the family caregivers puts them at risk for poor decision-making. This study will explore how AYA decision-making involvement influences transition readiness as an essential first step in developing better transition preparation processes for AYAs with SCD who face greater socioeconomic challenges, discrimination, and health disparities than other medically complex conditions (DeBaun & Telfair, 2012; Elander et al., 2003; Jenerette & Brewer, 2010; National Heart, 2018).

Aim 1: To explore the relationship between decision-making involvement and self-efficacy in AYAs with SCD.

RQ 1: Is there a correlation between decision-making involvement and self-efficacy in AYAs with SCD?

Aim 2: To explore the relationship between family caregiver's perception of decision-making involvement and self-efficacy for AYAs with SCD.

RQ 2: Is there a correlation between family caregiver's perception of decision-making involvement and self-efficacy in AYAs with SCD?

Aim 3: To predict transition readiness based upon AYA's decision-making involvement and self-efficacy after controlling for AYA and family characteristics.

RQ 3: After controlling for AYA and family characteristics, to what degree does the combination of AYA's decision-making involvement and self-efficacy predict transition readiness in AYAs with SCD?

This preliminary study is the first in a planned program of research to develop patient- and family-centered interventions to improve transition readiness in AYAs with SCD. Decisions about management of SCD do not occur in isolation but are made within the context of family and environment. This study will inform our understanding of the transition process to increase early identification of AYAs at high risk for poor transition in order to development future multi-component interventions to address the transition preparation and self-management capacity of this unique population.

Research Strategy

Background & Significance

Incidence and Impact of Sickle Cell Disease. SCD is the most common blood disorder in the US; it is a group of inherited disorders affecting red blood cells (National Heart, 2018). The incidence of SCD is 15.5 per 1,000 newborns (Ojodu et al., 2014).

Individuals with SCD produce an abnormal hemoglobin known as hemoglobin S, commonly known as sickle hemoglobin (Liprie, 1993). Unlike normal hemoglobin, hemoglobin S has the ability to turn into sickle shapes (National Heart, 2018). These malformed cells are inflexible and sticky leading to blockages in blood vessels, which in turn reduces or stops the flow of blood leading to deoxygenation of the nearby tissues (National Heart, 2018). This abnormality causes sudden severe pain also known as vaso-occlusive crises, or pain crises, which often require additional medical treatment and hospitalizations for effective pain management and resolution (National Heart, 2018). As children age, and as result of repeated pain crises, they often begin to experience chronic pain (National Heart, 2018). Over a person's lifetime, SCD causes organ damage to the brain, spleen, liver, lungs, kidneys, heart, bones, skin, penis, and joints (National Heart, 2018). Severe and life-threatening complications of SCD include splenic sequestration crises, aplastic crises, infections, acute chest syndrome, and strokes (National Heart, 2018).

Routine screening for SCD at birth, early management, and improved treatments have increased the lifespan of persons with SCD (National Heart, 2018). Treatment for SCD includes hydroxyurea therapy, iron chelation therapies, and transfusions; all of which require lifelong management to ensure effectiveness and reduce opportunities for sickle cell crisis (National Heart, 2018). Disease management is challenging for AYAs with SCD and recent increases in lifespan have created the need to transition increasing numbers of AYAs from pediatric healthcare to adult healthcare settings for continued medical management (Brown & Sobota, 2016; Gardner et al., 2016; Hauser & Dorn, 1999; Jordan, Swerdlow, & Coates, 2013; Quinn et al., 2010; Speller-Brown et al., 2015;

Tuchman, Slap, & Britto, 2008). This transition period is dangerous for chronically ill AYAs, but the complex nature of SCD management puts AYAs at greater risk as they transition their healthcare teams and assume more responsibility for their own disease management (Quinn et al., 2010).

Transition and Associated Risks. The transition from the pediatric healthcare setting to the adult healthcare setting at the surface level appears to be just a simple transfer from one healthcare team to another; yet, in reality, this transition is a complex process with multiple stakeholders including patients, family caregivers, and healthcare providers (American Academy of Pediatrics, American Academy of Family Practice, & American College of Physicians [AAP], 2002). AYAs include adolescents (ages 13-17) and young adults (ages 18-21). Recently, national guidelines have called for adolescents as young as 13 years to be included in transition preparation (AAP, 2002; Transition, 2018). Transition readiness can take years to develop and early intervention allows adequate time for adolescents to gain knowledge and independence in their care (AAP, 2002; Transition, 2018). Young adults are those ages 18-21 years and commonly continue to receive care from pediatric healthcare providers until the individual turns 21. It is important to note that once an individual turns 18 in the pediatric healthcare setting, they are considered an adult and must give consent for themselves. It does not necessarily mean that they have taken over control of disease management at home. National guidelines recommend that young adults have their first appointment with an adult healthcare provider prior to turning 22 (Transition, 2018).

There are stark differences between pediatric and adult healthcare management. It is a standard of practice to use anticipatory guidance as well as an advocacy focused

approach in pediatrics to encourage continuity of care. As an example, when parents do not bring their child to clinic and miss care, healthcare providers follow-up, and, if they cannot resolve the issue, they are obligated to report potential medical neglect. The adult healthcare setting takes a starkly different approach where the responsibility is placed upon the young adult to attend appointments and manage their own care, and there is little follow-up for missed care. Additionally, there is a lack of adult healthcare providers with adequate knowledge of SCD leading to chronic use of the emergency rooms for primary care which in turn leads to missed opportunities to prevent or mitigate severe complications (Kanter & Kruse-Jarres, 2013). Normal developmental changes in the individual and family structure occurring during this time put the AYAs at risk as they assume the responsibilities of adulthood and begin spending less time with the family. At this same time families and parents are adjusting to generally reduced roles in their child's life, specifically to less intense disease management support (Zhou, Roberts, Dhaliwal, & Della, 2016). However, AYAs may not be prepared intellectually and psychologically to assume responsibility for their disease. Despite national calls for improvement in transition preparation, little progress has been made to advance knowledge and practice in this area of AYA transition readiness (McManus et al., 2013).

Both long and short term effects of inadequate preparation for transitions from pediatric to adult care have been well documented in a wide variety of chronic diseases including inflammatory bowel disease, chronic kidney disease, congenital heart disease, type 1 diabetes, and epilepsy (de Silva & Fishman, 2014; Fenton, Ferris, Ko, Javalkar, & Hooper, 2015; Goossens et al., 2011; Helgeson et al., 2013; Pyatak et al., 2017; P. Ryan & Sawin, 2009; Stam, Hartman, Deurloo, Groothoff, & Grootenhuys, 2006). Shortly after

transition to the adult healthcare setting, adolescents with chronic disease are at risk for increased emergency room visits, hospitalizations, poor self-management, and increased depressive symptoms (Brousseau et al., 2010; Fenton et al., 2015; Pyatak et al., 2017). Specifically, in the SCD population, hospitalizations were higher for young adults after transition to the adult healthcare setting in comparison to adolescents, and they also tended to incur higher medical costs (Brousseau et al., 2010; Dickerson et al., 2012). Young adults post-transition received fewer transfusions which are critical to reducing complications from SCD such as strokes and acute chest syndrome (odds ratio: 0.80, $p=0.002$) (Blinder et al., 2013). After transition, the number of hydroxyurea prescriptions filled dropped for young adults from 0.35 prescriptions per patient per quarter at 18 to 0.26 prescriptions per quarter per patient at age 25, and this can leave young adults with SCD at risk for high rates of vaso-occlusive crises from their SCD and increased end organ damage (Blinder et al., 2013). Of all the findings regarding health outcomes post-transition, the most significant was a study that found that most of the deaths in a study of 940 AYAs with SCD occurred with a mean time of 1.8 years after transition to the adult healthcare setting (Quinn et al., 2010). These poor outcomes demonstrate the critical need for improvement of transition for adolescents and young adults with SCD.

The SCD population has factors unique from other chronic illness, with severe pain being the most difficult to manage. Because of their frequent exposure to pain and treatment for pain since childhood, individuals can have a well-developed and thorough understanding of what types of medications and doses work for their pain (Barakat, Patterson, Daniel, Dampier, & Outcomes, 2008; Elander et al., 2003; Reiss, Gibson, & Walker, 2005). This knowledge about their most effective pain management regimen can

cause healthcare providers who are less familiar with SCD to label them as drug-seeking, particularly if they are seeking care from emergency departments (Elander et al., 2003). This type of discrimination can cause delays in care and inadequate treatment (Elander et al., 2003). Furthermore, 60% of AYAs with SCD are insured through Medicaid, which leads to insurance coverage gaps after age 18 when they have to apply for their own coverage (Brousseau et al., 2010). These socioeconomic challenges and health disparities put this population at further risk for complications and death and heightens the necessity of further study into developing SCD-specific transition studies and interventions.

Current State of SCD Transition Research. With improvements in screening and management of SCD, the US life expectancy has dramatically increased from 14 years in 1973 to 40-60 years today (Elmariah et al., 2014). Knowledge of how to prepare AYAs for the transition has not kept up with the advances in medical management for SCD. There are a few known non-modifiable and modifiable factors that influence transition readiness in AYAs with SCD. Age and SCD severity are non-modifiable factors. It has been found that as the AYA ages, transition readiness increases ($p = 0.003$) (McPherson, Thaniel, & Minniti, 2009). However, those with increased frequency of use of acute care services have an inverse relationship to transition readiness; those with more severe SCD had significantly less interest in activities related to transition readiness ($p = 0.043$) (McPherson et al., 2009). There are other non-modifiable factors that may relate to transition readiness. These factors include SCD genotype, insurance type, family structure, and family caregiver's level of education. While these factors have received limited attention, it is likely these variables influence transition readiness. Insurance type is a simplified way of assessing financial status; however, AYAs with SCD on Medicaid

or other public insurances might have limited access to resources making it more challenging to develop transition readiness. Additionally, at the point of transition to adult care, AYAs on Medicaid may experience a loss of insurance, which reduces their ability to access important healthcare.

Family factors may influence the development of transition readiness as family structure could affect the type of support an AYA with SCD has during transition preparation. Those with more stable and consistent support may have more backing as they practice and develop skills essential to successful transition. Family members' level of education and support for the AYA's self-management may affect the development of transition readiness. It has been shown that parental health literacy is negatively related to child health outcomes (DeWalt & Hink, 2009). Though level of education of family caregivers has not been studied for its relationship to transition readiness, it is hypothesized that AYAs who have caregivers with high levels of education will have higher levels of transition readiness. Understanding these factors would enable healthcare providers to personalize transition preparation for AYAs with SCD.

Self-efficacy in SCD self-management is the confidence the AYA has in their ability to manage the pain they experience related to their disease (Edwards et al., 2000). This is the only modifiable factor which has been found to be positively associated with transition readiness in adolescents and young adults with SCD prior to transition (Treadwell et al., 2016). As the individual's confidence increases, their ability to self-manage their SCD and their transition readiness both increase (Treadwell et al., 2016). Self-efficacy is a modifiable factor related to transition readiness, but is also a desired outcome of transition preparation. Ideally as the AYA with SCD is aging out of the

pediatric healthcare setting, it would be expected that they have higher self-efficacy, or confidence in their SCD knowledge and self-management skills, and thus, higher levels of transition readiness.

This proposed study looks to assess AYA decision-making involvement, a novel factor which may be related to self-efficacy in SCD self-management and transition readiness. AYA decision-making involvement is the way in which AYAs take part in the decision-making process with their parents regarding the daily management of their disease (Miller & Drotar, 2006). For example, AYAs could be passive decision-making by only listening to their family caregivers when a decision about disease management is being made. AYAs who are actively involved in decision-making for their disease management may contribute their opinions or knowledge to the conversation with their family caregiver when a decision is being made. Increasing decision-making involvement during transition preparation has been shown to enhance self-efficacy and influence self-management behaviors such as adherence, but this has not been studied in AYAs with SCD (Liprie, 1993; Miller & Drotar, 2006; F. A. White, 1996). There have been no studies reported linking decision-making involvement and transition readiness in AYAs living with SCD. This is the gap that this study is designed to address. It is hypothesized in this study that AYA decision-making involvement will be related to AYA self-efficacy and transition readiness in AYAs with SCD. It is hypothesized that, as the AYA engages more in decision-making regarding their disease, they are gaining confidence in their knowledge and skills related to disease self-management (Miller & Drotar, 2006).

A unique approach proposed with this planned research is to study AYA-family caregiver dyads. Prior research utilizing AYAs and family caregiver dyads has shown

that there is a discrepancy between AYA and family caregivers' perspective of transition readiness in AYAs with SCD (Speller-Brown et al., 2015). There has been no work that describes factors related to transition readiness in the AYA and family caregiver dyad (Pyatak et al., 2017). AYAs with SCD may view their decision-making involvement and transition readiness differently than their family caregivers. As transition preparation involves the transfer of responsibilities from family caregivers to the AYA, research utilizing AYA and family caregiver dyads allows for the development of a comprehensive understanding of process variables and their relationship to transition readiness.

Theoretical Framework. The Individual and Family Self-Management Theory (IFSMT) will guide this study. It provides an explanation of how the development of transition readiness occurs in the context of a variety of risk and protective factors, which influence the AYA's involvement in health behaviors required for successful transition to the adult healthcare setting (P. Ryan & Sawin, 2009). IFSMT posits that the development of optimal knowledge and self-management is a combination of active and purposeful involvement by the AYA coupled with family caregiver support leading to shared responsibility for illness management (P. Ryan & Sawin, 2009). This theory expands upon prior work in the field of self-management by recognizing that both the individuals and families as a dyad influence outcomes (P. Ryan & Sawin, 2009). This theory has been used by other research on transition of youth with chronic disease (Pai et al., 2010). Studying the AYA and family dyad gives a more complete understanding of the potential influence of family functioning and individual characteristics (context variables) on health behaviors (process variables) and transition readiness (outcome variable) (P. Ryan

& Sawin, 2009). As there is limited understanding of the impact of non-modifiable factors on modifiable transition readiness factors, understanding these relationships will increase the likelihood of developing successful interventions which take into account family caregiver and AYA roles in transition readiness. This study proposes to address this gap by exploring the relationship of AYA and family caregiver context, process, and outcome factors that influence AYA transition to adult healthcare settings.

Innovation of the Proposed Study and Study Significance. This study is innovative because it studies a novel modifiable variable, decision-making involvement, and transition readiness utilizing both the perspective of AYAs and family caregivers. Decisions about care occur within family units at home; the proposed strategy will provide insight on the roles AYAs take in take in decision-making occurring at home (i.e. decision-making involvement) and the potential influence of decision-making involvement on the AYA’s development of transition readiness. Changes in transition readiness cannot be made by studying the AYA alone, isolated from the family caregiver

influence. In the literature surrounding transition preparation and SCD, only one study has examined dyadic influences, and this proposed study will shed more light on the complex interplay of processes between AYAs and their family caregivers (Speller-Brown et al., 2015). Prior research has provided knowledge of non-modifiable variables related to transition readiness. This will be the first study utilizing established theory to examine the impact of potentially modifiable processes used in decision making about SCD. By assessing dyadic decision-making involvement, this study will provide new knowledge that can be targeted for future interventions.

This study has significant public health implications because transition of AYAs with SCD is associated with increases in morbidity and mortality. SCD disproportionately impacts African Americans, who are already at risk for discrimination and health disparities (Barakat et al., 2008; Elander et al., 2003; Jenerette & Brewer, 2010; National Heart, 2018; Reiss et al., 2005). Furthermore, this study aligns with the goals of the National Institute of Nursing Research (NINR) and with the Healthy People 2020 goals. The NINR has made increasing quality of life one of its five main areas of focus and has asserted that self-management is a key component to meeting quality of life goals (Research, 2018). To develop transition readiness, AYAs must have adequate disease-specific knowledge and self-management skills needed for successful transition to the adult healthcare setting. Without understanding more about the complex interplay of factors and family influence, it is difficult to develop interventions that will be successful in improving AYAs with SCD transitions to adult healthcare settings.

Preliminary Studies at Children's National Medical Center. This study builds on previous work done in the area of transition readiness by Dr. Barbara Speller-Brown

and collaborative work between the investigator and Dr. Speller-Brown. Dr. Speller-Brown had previously studied the perceptions of transition readiness among the AYA-parent dyad in the SCD population. This study found that AYA age, the AYA's level of responsibility for disease self-management, and the level of parent involvement were significantly associated with transition readiness (Speller-Brown et al., 2015). The principal investigator from this study then collaborated with Dr. Speller-Brown on a descriptive study which described transition and disease knowledge of parents and youth with sickle cell disease who sought care from an urban comprehensive sickle cell clinic. This study identified that while knowledge of the disease was high in parents and youth with sickle cell disease, there was limited understanding and knowledge of steps necessary to performing self-management behaviors on both well and sick days in youth with sickle cell disease (Speller-Brown, Varty, Thaniel, & Jacobs, 2019). This experience offered the opportunity to develop a working relationship with Dr. Speller-Brown which will facilitate our partnership for this study. Dr. Speller-Brown and the investigator spent time discussing this study proposal and the challenges and strategies for carrying out this work at Children's National Medical Center. In addition, this prior research experience demonstrates the student's ability to successfully collaborate within a research team.

Approach

Study Design. The proposed study is a correlational design examining context and process variables, prediction on transition readiness in AYAs with SCD prior to transition to the adult healthcare setting. Context, process, and outcome variables will be

operationalized using validated measurement tools, documentation in the electronic medical record, and self-report when appropriate.

Setting. The setting for the study is Children's National Medical Center Comprehensive Sickle Cell Disease clinic which is an outpatient clinic in a large, urban pediatric hospital in Washington, District of Columbia (DC). The patients are primarily from zip codes in DC, Maryland, and Virginia. Members of the patient population are primarily African American and have Medicaid as their primary insurance; this is similar to the general SCD population (National Heart, 2018; Raphael et al., 2009). This is the largest pediatric SCD clinic in the US and sees over 1,400 SCD patients per year with ages from birth to 21. When patients turn 18, a majority of young adults begin attending a clinic run by healthcare providers whose focus is transition preparation. Transition must at least occur for the individual after they turn 21 years old due to reimbursement from insurance.

Sample. For this study, a convenient sample of all eligible subjects in the SCD clinic will be recruited. A power analysis was conducted, and it was determined that 42 family caregiver-child dyads are needed. The power analysis was conducted using G*Power utilizing a significance of 0.05 and 80% power. A total of 50 dyads will be recruited to account for attrition. AYAs will be stratified by two groups: 13-17 years of age and 18-21. The study is including adolescents as young as 13 because national guidelines suggest that transition preparation should begin in early adolescence to allow adequate time for knowledge and self-management skill development (AAP, 2002; Transition, 2018). Young adults are included as it is common practice for young adults to stay in the pediatric healthcare setting until insurance no longer covers their care. Fifty

patients and 50 caregivers from the SCD clinic will be recruited. The patients will be stratified equally into an adolescent and a young adult group. The groups help to ensure that age groups are evenly weighted, which is important because the AYAs are in varying developmental stages, and those 18 and older will have begun transition preparation which is a standard of care in the outpatient comprehensive SCD clinic. Approximately 450 patients in the clinic are between the ages of 13-21. The clinic treats patients from all levels of socioeconomic status, insurance types, and background, and this assures subject selection is equitable meaning all groups in the population of interest will have access.

Inclusion/Exclusion Criteria. There are two eligibility groups. For AYAs to be eligible, the AYA will need to have a diagnosis of SCD (any genotype), be between the ages of 13-21 at the time of entry into the study, and speak English as their primary language. All adolescents under the age of 18 will need to provide assent in addition to their family caregivers providing consent. For those 18 and older, consent will be required for both AYA and parent participation. Young adults who attend clinic alone will not be excluded. A separate protocol will be followed so that the study can be completed via a telephone call at a later, agreed upon time. For the family caregiver to be eligible, they will be required to have English as their primary language, be the primary family caregiver who manages the AYA's disease management, and provide consent to be included in the study. Family caregivers who do not participate in SCD management at home will be excluded.

Subject Recruitment and Retention. With the assistance of the healthcare team in the clinic, potential subjects who meet the inclusion criteria will be identified prior to their scheduled clinic visits during weekly meetings with members of the SCD team.

Participants will be recruited during regular clinic visits. The consent and the one-time data collection will occur during the same visit to reduce loss to follow-up. Loss to follow-up is a challenge in this population as they frequently do not have access to transportation and frequently change their telephone numbers and places of residence. Prior research in this population utilizing one-time survey administration during clinic visits has been effective and informed the development of this research strategy (Speller-Brown et al., 2015). Consent and assent will be obtained by the principal investigator. After consultation with a CNHS IRB analyst, I will submit a waiver of documentation of written consent because I will not be collecting any protected health information. This waiver is used when a written consent form would be the only link between the participant and the study data. I will employ procedures my CNHS mentor, Dr. Kathy Kelly has used in her previous research of child cancer communication. I will review and /or read the information sheet describing the study to both the AYA and the family caregiver. Participants will be given time to consider their participation and to review the information sheets. If they agree to participate, then their verbal agreement will be considered confirmation of consent and/or assent according to CNHS IRB policy and procedure regarding Waiver of Documentation of Informed Consent. Please see the copy of the IRBear study protocol for the specific steps. Participants and their family caregivers will be informed that consent and assent can be withdrawn at any time. Participants will receive a \$15 gift card at the end of completing their surveys to incentivize their participation.

Measures. All measures will be collected at a single time point. Variables will be derived from electronic health records, family caregiver report, or through the use of validated instruments as described in the table below.

Table 7

Description of Variables and Methods for Data Collection

Construct	Instrument/Source	Participant Role	Variable Operationalization
<i>Covariates - (Context Variables)</i>			
Frequency of use of acute care services	Electronic Medical Record	None	Number of emergency room visits or hospital stays related to SCD in prior year
SCD genotype	Self-report	None	Type of SCD genotype
Gender	Self-report	None	Gender listed in the electronic medical record
Insurance Type	Self-report	None	Insurance type
AYA's Age	Self-report	None	Age in years
Family Structure	Genogram	AYA self-report	Genogram
Family Members Level of Education	Self-report	Family caregiver self-report	Highest level of education obtained by each family member and noted on genogram
<i>Independent Variables - (Process Variables)</i>			Psychometric Properties
Self-efficacy	Sickle Cell Self-Efficacy Scale	AYA completion of tool	<ul style="list-style-type: none"> • Internal consistency (over all Cronbach's alpha = 0.89)¹² • Divergent, construct, and predictive validity¹²
Decision-making involvement	Decision-Making Involvement Scale (DMIS)	Family caregiver completion of parent-DMIS version and AYA completion of AYA-DMIS version	<ul style="list-style-type: none"> • Internal consistency (range of Cronbach's alpha from 0.71-0.91)³⁴ • Temporal and construct validity³⁴
<i>Dependent Variable - (Outcomes Variable)</i>			
Transition readiness	Readiness to Transition Questionnaire (RTQ)	Family caregiver completion of parent-RTQ version and AYA completion of AYA-RTQ version	<ul style="list-style-type: none"> • Internal consistency (range of Cronbach's alpha from 0.79-0.94)¹⁷ • Inter-rater reliability¹⁷ • Construct validity¹⁷

Context Variables. Frequency of use of acute care services, SCD genotype, gender, insurance type, AYA's age, family structure, and family members' levels of education will all be assessed as covariates because IFSMT asserts that disease condition factors, environment or financial factors, and family factors can influence process variables in the individual (P. Ryan & Sawin, 2009). Frequency of use of acute care services and AYA's age have been studied previously; however, SCD genotype, family structure, and family caregiver level of education have received limited attention.

Frequency of use of acute care services will be assessed as a continuous variable by emergency room visits or hospitalizations stays related to SCD in the prior year. SCD genotype will be categorized into the following types: HbSS, HbSC, HbS β + Thalassemia, and HbS β 0 Thalassemia. Insurance coverage type will be categorized by the following types: private insurance, private pay, public insurance (Medicaid), or uninsured. Family structure will be determined by asking the AYA to describe their family while the PI constructs a genogram reflecting the AYA's description. Wright and Leahey (2013) defined family as "family is who they say they are," so constructing the genogram will allow for the individual to provide the PI with the details of their family structure as they view them (Wright & Leahey, 2013). The PI will elicit feedback on the genogram from the AYA to ensure that it accurately depicts their family structure. Family caregivers' education level will be categorized by the following types: 12th grade or less, high school or GED, some college or technical training, college graduate, or graduate school.

Process Variables. Beliefs and self-regulation are related to self-management outcomes according to IFSMT (P. Ryan & Sawin, 2009). Self-efficacy in SCD self-management will be measured utilizing the validated Sickle Cell Self-Efficacy Scale

(SCSES) consisting of 9 items measuring an individual's perception of their ability to manage SCD (Edwards et al., 2000). It uses a Likert scale which ranges from "Not at all sure" to "Very sure" for scoring with higher scores indicating a higher the level of self-efficacy (Edwards et al., 2000). Divergent, construct, and predictive validity was established for this measurement tools in prior work with AYAs living with SCD (Edwards et al., 2000). The tool demonstrated strong internal consistency as demonstrated above in the table (Edwards et al., 2000). Self-regulation will be measured by assessing the decision-making involvement of AYAs with SCD using the Decision-Making Involvement Scale; a validated 30-item tool which has 5 subscales assessing the following behaviors: asking for advice, expressing an opinion, asking for an opinion, expressing advice, and negotiation and brainstorming (Miller & Harris, 2012). For each item, the family caregiver and the AYA are asked to use a Likert scale ("not at all", "a little bit", "quite a bit", and "a lot") to rate how the AYA participates in the decision-making process. Each subscale score can range from 1-4, and higher scores demonstrate more of that behavior or type of involvement (Miller & Harris, 2012). Temporal and construct validity were established for this measurement tool in prior work with AYAs living with chronic disease (specifically, diabetes mellitus) (Miller & Harris, 2012). The tool demonstrated strong internal consistency as demonstrated above in the table (Miller & Harris, 2012).

Outcome Variables. The outcome of interest is transition readiness of the AYA. Transition readiness is a measurement of the preparedness of the AYA to make the transfer to the adult healthcare setting. The development of transition readiness engages AYAs in health behaviors to self-manage their SCD including knowledge of disease,

medication management, communication, and utilization of healthcare resources. The Readiness for Transition Questionnaire (RTQ) will be utilized to determine the level of participation of the AYA in healthcare behaviors essential for managing their disease; the RTQ has been validated in the SCD population (Gilleland et al., 2012; Speller-Brown et al., 2015). The RTQ has an AYA self-report scale and family caregiver scale which requires the family caregiver to report on their perception of the AYA's transition readiness (Gilleland et al., 2012; Speller-Brown et al., 2015). For each item, the family caregiver and the AYA are asked to use a Likert scale ("not responsible at all", "sometimes responsible", "often responsible", "almost always responsible") to rate how the AYA participates in health care behaviors (Gilleland et al., 2012). The scores are added with higher scores indicating higher levels of transition readiness (Gilleland et al., 2012). Inter-rater reliability and construct validity were established for this measurement in prior work with AYAs living with chronic disease (specifically, chronic kidney disease) (Gilleland et al., 2012). The tool demonstrated strong internal consistency as demonstrated above in the table (Gilleland et al., 2012).

Data Management and Analysis Plan. All data collected during the study will be stored in a secure, password-protected laptop computer. All data on paper forms and the external hard drive with backup data will be stored in locked file cabinet to which only the PI has access. All the data collected from both the medical records and the paper and pencil forms will be entered into a REDCap file so that it can be analyzed using Statistical Package for Social Sciences (SPSS) Version 25. In the event of missing data, the PI will impute the missing data points using an appropriate statistical algorithm based upon the type of missing data in consultation with the statistician. Analysis will begin

with generating descriptive statistics about frequency of use of acute care services, SCD genotype, gender, socioeconomic status, age, family structure, family member level of education, SCSES scores, DMIS scores, and RTQ scores. Then an appropriate analysis will be run to answer each research question. Significance will be set at $p < 0.05$.

Table 8

Description of Analysis Method to Address Study Aims

Aim Number	Level of Measurement for Data	Analysis Method	Assumptions to Test for Analysis Method	Alternative Analysis Method
Aim 1	<ul style="list-style-type: none"> • AYA's Decision-Making Involvement score - continuous • Sickle Cell Self-Efficacy Score – continuous 	Pearson product-moment correlation analysis	<ul style="list-style-type: none"> • Normal Distribution (assessment of distribution of SCSES scores) • Linearity (Shapiro-Wilk and Kolmogorov-Smirnov tests) • Homoscedasticity (assessment of residuals and distribution around fitted regression line) 	Spearman's correlation
Aim 2	<ul style="list-style-type: none"> • Family caregiver's Decision-Making Involvement score - continuous • Sickle Cell Self-Efficacy Score - continuous 	Pearson product-moment correlation analysis	<ul style="list-style-type: none"> • Normal Distribution (assessment of distribution of SCSES scores) • Linearity (Shapiro-Wilk and Kolmogorov-Smirnov tests) • Homoscedasticity (assessment of residuals and distribution around fitted regression line) 	Spearman's correlation
Aim 3	<ul style="list-style-type: none"> • AYA and Family caregiver Readiness to Transition score – continuous • AYA's Decision-Making Involvement score - continuous • Family/ caregiver's Decision-Making Involvement score - continuous 	2 Multiple linear regression	<ul style="list-style-type: none"> • Normality (assessment of distribution of RTQ scores prior to conducting multiple linear regression analysis and assessment of normal P-P plot after analysis) • Linearity and homogeneity (assessment of plot of standardized predicted values against standardized residuals) • Outliers (assessing residuals using standard deviations) • Multicollinearity (assessment of VIF values) 	N/A

Aim 1: The Pearson product-moment correlation will determine if there is a linear relationship between AYA's perception of their decision-making involvement and their

self-efficacy, and, if there is a relationship, it will also provide a description of the direction of that relationship.

Aim 2: The Pearson product-moment correlation will determine if there is a linear relationship between family caregiver's perception of the AYA's participation in decision-making involvement and AYA's self-efficacy, and, if there is a relationship, it will also provide a description of the direction of that relationship.

Aim 3: Two multiple linear regression analysis will be conducted to answer research question 3, because the RTQ scores are continuous and can control for covariates. One linear regression will study the relationship between the independent variables and the AYA's RTQ scores, and one linear regression will study the relationship between the independent variables and the family caregiver's RTQ scores.

Type I error will be reduced by using an alpha level of 0.05. The Type II error will be reduced by setting beta at 0.80 for the study and by recruiting at least as many subjects as determined from the power analysis or greater. Missing data will be examined in consultation with the statistician to develop a plan for how best approach the analysis.

End Point to Study. The end point of this study will be the development of a description of the factors associated with transition readiness in adolescents and young adults living with sickle cell disease. The researcher designed this study to provide a conceptual understanding of how the decision-making involvement regarding disease management at home for AYAs living with SCDS influences their readiness to transition. I will use this framework to inform my future research of AYA transition in youth with chronic illness, which will include interventions targeting highly predictive factors influencing AYA transition readiness.

Limitations. As with all research, there are limitations. First, there may be history bias as AYAs may have been exposed to varying levels of education regarding transition preparation and the need to develop self-management. A threat to internal validity includes the use of self-report tools. The AYA or family caregivers may over or under-report the AYA's decision-making involvement or transition readiness. By using self-report measurement tools that also have family caregiver self-report, the PI is able to gain perspective from multiple points of view regarding decision-making involvement and transition readiness. One threat to external validity includes selection bias. The participants for this research study will be recruited from a large, urban pediatric hospital and the results may not be generalizable to rural populations.

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CHAPTER FIVE

DECISION-MAKING INVOLVEMENT, SELF-EFFICACY, AND THE RELATIONSHIP TO TRANSITION READINESS IN YOUTH WITH SICKLE CELL DISEASE

*As the primary author, my colleagues and I plan to submit for publication to the Journal
of Pediatric Nursing*

Abstract

Introduction: Transition to adult healthcare is a critical time for adolescents and young adults (AYA) with sickle cell disease (SCD). Preparation for transition is important to reducing morbidity and mortality risks. The aim of this correlational, cross-sectional study was to examine the relationships between decision-making involvement, self-efficacy of SCD self-management, AYA healthcare responsibility, and transition readiness in AYA with SCD prior to transition to adult healthcare settings.

Methods: This descriptive, cross-sectional study was conducted with 50 family caregiver-AYA dyads who were receiving care from a large urban comprehensive sickle cell clinic in Washington, DC between October 2019 and February 2020. The data were collected using the Decision-Making Involvement Scale, the Sickle Cell Self-Efficacy Scale, the Readiness to Transition Questionnaire, a demographics survey, and the electronic medical record. Data were analyzed using Pearson product-moment correlations and multiple linear regression.

Results: This study found that while higher levels of expressive behaviors such as sharing opinions and ideas in decision-making were associated with higher levels of AYA healthcare responsibility, this type of behavior was inversely associated with feelings of

transition readiness. Self-efficacy was positively associated with transition readiness but inversely related to AYA healthcare responsibility. Parent involvement was negatively associated with AYA healthcare responsibility.

Discussion: Future research interventions that facilitate active AYA involvement in decision-making regarding their disease, increase self-efficacy, and safely reduce parent involvement may positively influence their capacity for self-management. While increasing AYA decision-making involvement may improve AYA healthcare responsibility, it may not reduce the barriers of feeling unprepared for the transition to adult healthcare.

Key words: transition readiness, transition, decision-making involvement, self-efficacy, sickle cell disease

With the use of routine screening at birth and improvements in treatment, the life expectancy for individuals living with SCD in the US has dramatically increased from 14 years in the 1970s to 40-60 years today (Elmariah et al., 2014). Youth with SCD age out of the pediatric health care setting in their early twenties, making the transition to adult healthcare critical to assuring continuity of care for young adults with SCD (Brown & Sobota, 2016; Gardner et al., 2016; Hauser & Dorn, 1999; Jordan, Swerdlow, & Coates, 2013; Quinn, Rogers, McCavit, & Buchanan, 2010; Tuchman, Slap, & Britto, 2008). Developing transition preparation processes to increase successful transitions for AYAs living with SCD has become a priority as multimorbidity increases with age (Ballas, 2018; Van Tuijn, Schimmel, Van Beers, Nur, & Biemond, 2017). After AYA transition to adult healthcare there is an alarming increase in morbidity and mortality (Blinder et al., 2013; Brousseau, Owens, Mosso, Panepinto, & Steiner, 2010; Lanzkron, Carroll, & Haywood, 2013; Paulukonis et al., 2014; Quinn et al., 2010). After transition, young adults have the highest rates of frequent acute care visits related to SCD complications (Dickerson, Klima, Rhodes, & O'Brien, 2012; Quinn et al., 2010). The most concerning risk related to poor transition is the sudden increase in mortality (Blinder et al., 2013). With over 93% of individuals with SCD living into adulthood, addressing transition is essential to reduce poor health outcomes (Quinn et al., 2010).

Increasing transition readiness has been proposed as a mechanism for improving health outcomes for AYAs with SCD after transition. To design interventions which address this, it is important to understand the non-modifiable and modifiable factors that influence transition readiness in AYAs with SCD (Varty & Popejoy, 2020). Only a few non-modifiable factors associated with transition readiness have been identified. Notably,

transition readiness increases as age increases in AYA with SCD (McPherson, Thaniel, & Minniti, 2009). Frequency of acute care services use, a measure of disease severity, has been found to be negatively related to transition readiness. Youth reporting higher use of acute care services are significantly less interested in health behaviors associated with transition readiness (McPherson et al., 2009). The only modifiable factor reported to have associations with transition readiness in AYAs with SCD is self-efficacy (Treadwell et al., 2016). Self-efficacy has been explored across other chronic diseases and similar findings regarding transition readiness have been found (Carlsen et al., 2017; Eaton et al., 2017; Gumidyala et al., 2018; Pakdeeprom, In-Iw, Chintanadilok, Wichiencharoen, & Manaboriboon, 2012; Sawicki, Kelemen, & Weitzman, 2014; Stewart et al., 2017; Uzark et al., 2015). As AYAs' self-efficacy increased so has their transition readiness (Treadwell et al., 2016). Since self-efficacy can be modified, it could be used as an expected outcome of preparing AYA for transition.

Guided by the Individual and Family Self-Management Theory, in this study we proposed to assess AYA decision-making involvement, a novel factor which may be associated with self-efficacy and transition readiness. IFSMT posits that the development of optimal knowledge and self-management is a combination of active and purposeful involvement by the AYA coupled with family caregiver support leading to shared responsibility for illness management (Ryan & Sawin, 2009). This theory expands upon prior work in the field of self-management by recognizing that both the individual and family as a dyad influence outcomes (Ryan & Sawin, 2009). This theory has been used by other research on transition of youth with chronic disease (Pai et al., 2010). Studying the AYA and family dyad gives a more complete understanding of the potential influence

of family functioning and individual characteristics (context variables) on health behaviors (process variables) and transition readiness (outcome variable) (Ryan & Sawin, 2009). As there is limited understanding of the impact of non-modifiable factors on modifiable transition readiness factors, understanding these relationships will increase the likelihood of developing successful interventions which take into account family caregiver and AYA roles in transition readiness. In this study we proposed to address this gap by exploring the relationships between AYA and family caregiver factors (context factors), self-efficacy, decision-making involvement (process factors), and transition readiness (outcome factors) that influence AYA transition to adult healthcare settings.

For this study, AYA decision-making involvement was defined as the contributions AYAs make to the decision-making process regarding their disease management (Miller & Drotar, 2006). These contributions could come in many forms such as active-listening, asking questions, offering ideas, sharing opinions or providing information (Miller & Drotar, 2006). Prior research found increasing decision-making involvement to be positively related to self-efficacy and self-management behaviors such as adherence, but this relationship has not been studied in AYAs with SCD (Liprie, 1993; Miller & Drotar, 2006; White, 1996). In addition, no studies have been conducted to assess the relationship between decision-making involvement and transition readiness in AYAs living with SCD. The purpose of this cross-sectional descriptive study was to examine the relationships between AYA decision-making involvement, self-efficacy of sickle cell disease self-management, and transition readiness in AYA with SCD prior to transition to adult healthcare settings. Aim one and two explored the relationships between AYA and family caregiver's perception of AYA decision-making involvement

and self-efficacy in AYAs with SCD. Aim three looked to predict AYA transition readiness based upon AYA's decision-making involvement and self-efficacy levels after controlling for AYA and family characteristics.

Methods

Prior to participant recruitment and data collection, institutional board review (IRB) approval was obtained from both the Children's National Medical Center's IRB and University of Missouri-Columbia Health Sciences IRB. The study was conducted at the Children's National Hospital Comprehensive Sickle Cell Disease clinic which is an outpatient clinic in a large, urban pediatric hospital in Washington, DC. In 2014, the clinic began routinely implementing transition preparation and education interventions for patients 18 and older until they transitioned to adult healthcare. In addition, the healthcare providers actively began to directly engage their patients in conversation and placed emphasis on encouraging the emerging adolescent to play a dominant role in understanding and managing their treatment plan during routine clinic care.

Study Sample

The majority of patients from this clinic lived in the District of Columbia, Maryland, and Virginia. Members of the patient population were primarily African American and had Medicaid as their primary insurance; this is similar to the general SCD population (National Heart, Lung, and Blood Institute, 2018; Raphael et al., 2009). Power analysis was conducted utilizing G*power, and it was determined that 42 family caregiver-child dyads were needed (Faul, Erdfelder, Buchner, & Lang, 2009). A total of 50 dyads were recruited to participate in the study. AYAs were stratified evenly by two groups: 13-17 years of age (adolescents) and 18-21 (young adults). This was important

because AYAs are in varying developmental stages, and those 18 and older would have begun transition preparation which is a standard of care in the outpatient comprehensive SCD clinic utilized for recruitment. The study included adolescents as young as 13 because national guidelines suggest that transition preparation should begin in early adolescence to allow adequate time for AYAs to attain knowledge and develop self-management skill (American Academy of Pediatrics, American Academy of Family Practice, & American College of Physicians [AAP], 2002; Got Transition, 2018). The study included young adults as old as 21 as it is common practice for young adults to stay in pediatric healthcare settings until they age out because of hospital policies or changes to insurance coverage. A total of 25 adolescents-family caregiver dyads and 25 young adult-family caregiver dyads were recruited to ensure that the age groups were evenly weighted.

There were two eligibility groups. For AYAs to be eligible, they needed to have a diagnosis of SCD (any genotype), be between the ages of 13-21 at the time of entry into the study, and speak English as their primary language. All adolescents under the age of 18 were required to provide assent in addition to their family caregivers providing consent. For those 18 and older, consent was required for both AYA and parent participation. For the family caregiver to be eligible, they had to speak English as their primary language and be the primary family caregiver who manages the AYA's disease management.

Procedures

With the assistance of the clinic healthcare team, participants were recruited during regular clinic visits. The consent and the one-time data collection occurred during

the same visit to reduce loss to follow-up. Data collection began with the PI and AYA drawing a genogram. These genograms identified family members who were the AYA's primary family caregivers. This information was then used to determine if the family member present was eligible to participate in the study. If the family caregiver who was considered the primary caregiver was not present at the appointment, then the study was completed over the phone with the AYA and family caregiver at an agreed upon time.

The study began with asking the dyad to describe a decision which they had made in the last two weeks. The dyad was then asked to identify the issue for which a decision needed to be made, whether or not a decision was made, and who made the decision (could be a joint decision). Then AYA and family caregiver were then asked to complete the Decision-Making Involvement Scale (DMIS) and Readiness to Transition Questionnaire (RTQ). AYA's were also given the Sickle Cell Self-Efficacy Scale (SCSES) survey to complete. Finally, demographic surveys were also completed with the AYA and family caregiver. Participants who completed the study received a gift card. The PI then collected the frequency of use of acute care services from the medical record. All data were entered into a REDCap survey for analysis (Harris et al., 2009).

Measures

All measures were collected at a single time point with 100% of the participants fully completing all instruments. Variables were derived from electronic health records, AYA self-report, family caregiver self-report, or through the use of validated instruments. Details are provided in Table 9.

Statistical Analyses

The data was analyzed using IBM SPSS Statistics 25.0 statistical software. Descriptive statistics were generated for AYA age, AYA gender, AYA SCD type, frequency of use of acute care services in prior year, family caregiver gender, family caregiver education level, number of reported primary family caregivers, family structure, and insurance type. For aims one and two, a Pearson product-moment correlation was conducted since the SCSES was normally distributed. For aim three, four linear regressions examined the relationship between (a) the independent variables (self-efficacy and decision-making involvement), (b) covariates and (c) the dependent variables (AYA self-reported perception transition readiness, AYA self-reported responsibility for tasks necessary for transition readiness, family caregiver perception of AYA's transition readiness and family caregiver perception of AYA's responsibility for tasks necessary for transition readiness). Significance was set at $p < 0.05$.

Results

Sample Characteristics

AYA and family caregiver demographic characteristics are found in Table 10. Of 60 dyads that were approached for the study, 50 (83%) dyads successfully completed the study. One dyad did not complete the study because the AYA had a neurocognitive disorder making it difficult to complete the surveys. Five dyads declined to participate due to time constraints. Four dyads did not complete the study, because the AYAs were not feeling well enough or were being admitted to the hospital for pain crisis management. The majority of participants were female in both AYA ($n = 29$, 58%) and family caregivers ($n = 44$, 88%). Additionally, 43 (86%) AYAs reported having one

primary family caregiver while 7 (14%) AYAs reported having two or more primary family caregivers.

Correlates between AYA Decision-Making Involvement Subscales (Child Express and Child Seek) and AYA Self-Efficacy

Pearson product-moment correlations coefficients were used to assess the relationship between two DMIS subscales and AYA self-efficacy to address aim one and two. DMIS subscales Child Express and Child Seek measure the degree to which AYA take active roles in decision-making. AYA self-efficacy measures the AYA's perception of their ability to manage their SCD. The results can be found in Table 11. AYAs reporting greater Child Express were significantly associated with greater AYA self-efficacy. There were no significant relationships between AYA report of Child Seek, family caregiver report of Child Express, or family caregiver report Child Seek and AYA self-efficacy.

Predictors of Perceived Transition Readiness and Health Responsibility

Four linear regression models were used to assess the relationship between (a) DMIS subscale scores, (b) SCSES scores, (c) AYA/family caregiver characteristics and (c) RTQ subscale scores (AYA and family caregiver report of AYA transition readiness and AYA healthcare responsibility) to address aim three. Table 12 shows variables which were found to be statistically significant in predicting AYA self-reported transition readiness.

The multiple linear regression analysis explained 65.9% of the variance in AYA's self-reported transition readiness. The results from the linear regression showed that higher levels of AYA's self-reported transition readiness were associated with being

female ($\beta = -0.276, p = 0.014$), having a lower age ($\beta = -0.356, p = 0.033$), and having a diagnosis of SCD type HbSS, ($\beta = 0.303, p = 0.007$). AYA transition readiness was also significantly associated with having higher scores on the DMIS Child Seek subscale ($\beta = 0.374, p = 0.020$), having lower scores on the DMIS AYA reported Child Express subscale ($\beta = -0.454, p = 0.009$), and lower scores on the DMIS AYA reported Joint Decision subscale ($\beta = -0.388, p = 0.006$). AYA transition readiness was also significantly associated with higher self-efficacy scores ($\beta = 0.304, p = 0.030$), higher AYA self-reported healthcare responsibility ($\beta = 0.704, p = 0.000$), and lower family caregiver self-reported parent involvement ($\beta = -0.503, p = 0.001$). Finally, AYA transition readiness was significantly associated with having public or dual private and public insurance ($\beta = -0.276, p = 0.014$).

The multiple linear regression analysis explained 67.4% of the variance in AYA's self-reported healthcare responsibility. Variables which were found to be statistically significant in predicting AYA self-reported healthcare responsibility are found in Table 13. The linear regression found that higher levels of AYA's self-reported healthcare responsibility were associated with older AYA age ($\beta = 0.106, p = 0.001$). It was also associated with higher scores on the DMIS AYA reported Child Express subscale ($\beta = 0.310, p = 0.007$) and higher scores on the DMIS AYA reported Joint Decision subscale ($\beta = 0.236, p = 0.017$). Higher AYA's self-reported healthcare responsibility was associated with lower self-efficacy scores ($\beta = -0.273, p = 0.018$), lower AYA reported parent involvement ($\beta = -0.317, p = 0.008$), and higher AYA self-reported transition readiness ($\beta = 0.408, p = 0.000$).

The multiple linear regression analysis explained 75% of the variance in family caregiver perceived AYA transition readiness. Variables found to be statistically significant in predicting family caregiver's perception of AYA transition readiness are found in the Table 14. The linear regression showed that higher levels of family caregivers perceived AYA transition readiness were associated with higher scores on the DMIS family caregiver reported Child Express subscale ($\beta = 0.232, p = 0.009$), higher AYA self-reported transition readiness ($\beta = 0.283, p = 0.002$), and higher family caregiver reported AYA healthcare responsibility ($\beta = 0.674, p = 0.000$).

The multiple linear regression analysis explained 79% of the variance in family caregiver perceived AYA healthcare responsibility. Variables found to be statistically significant in predicting family caregiver's perception of AYA healthcare responsibility are found in Table 15. The results from the linear regression showed that higher levels of family caregivers perceived AYA healthcare responsibility were associated with having a SCD type other than HbSS ($\beta = -0.152, p = 0.049$). It was also associated with higher scores on the DMIS family caregiver reported Joint Decision subscale ($\beta = 0.193, p = 0.010$). Family caregiver perceived AYA healthcare responsibility was associated with lower family caregiver self-reported parent involvement ($\beta = -0.295, p = 0.003$) and higher family caregiver reported AYA transition readiness ($\beta = 0.607, p = 0.000$).

Discussion

It is widely accepted that developing transition readiness is a vital first step in preparing AYA for transition to adult healthcare (AAP, 2002). Currently assessment for transition readiness occurs using established tools, yet little is actually known about what factors are associated with transition readiness in AYA living with SCD or how to

positively influence transition readiness in this population (Varty & Popejoy, 2020; Zhang, Ho, & Kennedy, 2014). This study specifically evaluated the relationship of potentially modifiable variables (decision-making involvement and self-efficacy) to transition readiness in AYAs living at home.

This study established that there is a positive relationship between AYA decision-making involvement and AYA healthcare responsibility, and that there is a negative relationship between parent involvement levels and AYA healthcare responsibility. With these findings it is suggestive that as AYAs take a greater role in decision-making they are possibly learning more about how to make good decisions and are also potentially get more opportunities to practice managing their SCD independently. With decision-making conversations there is a potential for the AYA to not only assert desire for independence but there is also chance for family caregivers to hear that their AYA would like to take on more responsibility. This may help the family caregiver to gradually shift responsibility from the family caregiver to AYA over time. This study identified that parental involvement had a negative impact on AYA's healthcare responsibility. Therefore, designing interventions to systematically reduce parent involvement may provide AYA with the opportunities to learn how to perform essential disease self-management behaviors such as managing clinic appointments, taking medications independently, and directing the conversations with healthcare providers regarding their treatment plan.

This study also reaffirmed findings from previous work that there is a positive relationship between AYA self-efficacy and AYA expressing behaviors in decision-making involvement (Miller, 2017), and that there is a positive relationship between AYA self-efficacy and AYA self-reported transition readiness (Treadwell et al., 2016).

Additionally, this study reaffirmed the positive relationship between being female and transition readiness (Treadwell et al., 2016), and the positive relationship between AYA age and AYA self-reported healthcare responsibility (Speller-Brown et al., 2015). Replication of these findings reassures their importance for inclusion in intervention development.

These findings support the development of interventions designed to increase AYA self-efficacy and AYA decision-making involvement while also working with parents to systematically reduce their involvement. It is imperative to assess how factors such as transition readiness, AYA healthcare responsibility, self-efficacy, decision-making involvement, and parent involvement predict outcomes after the AYA transition to adult healthcare. The science of transition readiness in AYA living with SCD cannot move forward without understanding how transition readiness and its related factors influences future outcomes such as frequency of hospitalizations, adherence, appointment attendance, frequency of complications, and young adult satisfaction with their new healthcare providers.

While incorporating AYA self-efficacy, AYA decision-making involvement, and parent involvement variables into interventions to increase transition readiness, it is also important to remember that development of transition readiness occurs primarily in the home environment. Routine clinic visits are time-limited and it is not possible for healthcare providers to address every facet of transition readiness or to increase AYA healthcare responsibility. Interventions designed to address decision-making involvement, self-efficacy, and parent involvement, will need to target family approaches to disease management in the home. Interventions should be targeted to developing AYA

self-management while simultaneously supporting family caregivers to slowly relinquish their control over responsibility for SCD management. Healthcare providers need to assist the family caregiver in encouraging their AYA to take part in decision-making so the AYA can gain experience with critical thinking and evaluating choices while still in the home environment. Furthermore, interventions for the AYA would benefit from a clear understanding that transition preparation in the pediatric setting is just the beginning of the transition process.

Longitudinal studies which follow cohorts of AYA living with SCD from the beginning of transition preparation to an extended period post-transition are needed to understand how these factors affect long term outcomes. Self-efficacy, AYA decision-making involvement, parent involvement, AYA healthcare responsibility, and transition readiness should be evaluated for their ability to predict and influence health outcomes after the AYA living with SCD has transitioned to the adult healthcare setting. In addition preparing the AYA prior to transition is just one component of successful transition. Not all AYAs will have been successfully prepared prior to transition, so interventions must be developed to assist them after transition and recognize those who may be at risk for poor health outcomes. Adult healthcare settings need to be engaged with pediatric healthcare settings to develop coordinated transition programs which may assist with ensuring those who lack readiness are given appropriate assistance to succeed in the adult healthcare setting.

The study identified that counter-intuitively AYAs who expressed themselves more during the decision-making process by offering opinions or ideas, were less likely to feel ready to transition. While the reasoning for this finding is unclear, we think there

is a possibility that this finding was influenced by the unique clinic setting where the study occurred. This clinic had implemented systematic transition education for all young adults, and the structure of clinic visits for AYAs were designed to actively engage them in discussions about disease management. Potentially as the participants were exposed to more information about transition and the changes they may experience in the future, the less confident they felt in their personal readiness for the transfer. From prior qualitative work, AYAs described many negative feelings associated with transition such as anxiety or apprehension due to the change in environments and healthcare teams (Bemrich-Stolz, Halanych, Howard, Hilliard, & Lebensburger, 2015; Porter, Graff, Lopez, & Hankins, 2014). Because AYAs in this clinic were informed years before transition about the impending change in healthcare teams, AYA may have developed concerns about leaving healthcare providers they have known for most of their childhood (Varty, Speller-Brown, Phillips, & Kelly, In Review). Future qualitative research should be conducted to better understand this unexpected response. Perhaps these findings are normative because the AYA has a good understanding about the changes that are coming. If so, additional counseling could be offered to help mitigate these negative feelings or interventions that bolster the AYA's confidence in their ability to assume responsibility for their care could be developed. Supportive counseling interventions designed to help the AYA voice their concerns as they prepare for transition and to practice needed skills might also be beneficial. These research findings must be explored in the future.

Another factor which should be considered is how the types of SCD and the potential experiences of living with specific types of SCD impact transition. A positive relationship between having the SCD type HgSS and AYA self-reported feelings of

transition readiness. Potentially this is a result of having more experience interacting with the healthcare system as they have more complications and hospitalizations. Time spend managing and experiencing complications, may have increased their confidence in feeling that they could handle the transfer to adult healthcare. It is also important to note though that this study found a negative relationship between the AYA having HgSS and their family caregiver's perception of the AYA healthcare responsibility. Potentially AYAs living with HgSS tend to have more severe disease and family caregivers may have had concerns about their AYA being independent and having the capacity to take full responsibility for their complex health condition. Moreover, if the AYA faltered in their self-management they were more likely to experience complications and family caregivers may have wanted to avoid the development of disease complications. Therefore, they remained more hands-on to ensure their AYA's wellbeing leading to having lower healthcare responsibility levels.

Limitations

This study had several limitations. First, there may have been bias introduced because AYAs were exposed to education about transition preparation and the need to develop self-management. The use of self-report tools may have threatened internal validity by over or under-reported the AYA's self-efficacy, decision-making involvement, or transition readiness. Selection bias was also a potential threat to external validity. The participants in this study were recruited from a large, urban pediatric hospital that had an established transition to adult healthcare education program. Therefore, the results may not be generalizable to AYAs with SCD who are cared for in programs without such programs or who live in rural settings. In addition, participants who did not show up or

cancelled their clinic visits were not included in the study, so these results may not be generalizable to those who frequently miss routine care.

Conclusions

This study assessed AYA decision-making involvement and self-efficacy for their relationship to AYA healthcare responsibility and transition readiness in AYA with SCD prior to transition. It was found that higher levels of expressive behaviors such as sharing opinions and ideas were associated with higher levels of AYA healthcare responsibility, but inversely associated with feelings of transition readiness. Future research interventions that facilitate active AYA involvement in decision-making about disease management may increase self-efficacy, and safely reduce parent involvement may positively influence their capacity for self-management. While increasing AYA decision-making involvement may improve AYA healthcare responsibility overall, it may not reduce the barriers of feeling unprepared for the transition to adult healthcare. Additional interventions must address this aspect of transition preparation for adult healthcare.

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Table 9

Description of Variables and Measures

Construct	Instrument/ Source	Participant Role	Variable Operationalization
<i>Covariates - (Context Variables)</i>			
Frequency of use of acute care services	Electronic medical record	None	Number of emergency room visits or hospital stays related to SCD in prior year
SCD genotype	Demographic survey	AYA self-report	Type of SCD genotype (e.g. HbSS, HbSC, HbS β + Thalassemia, HbS β 0 Thalassemia)
Gender	Demographic survey	AYA self-report	Gender listed in the electronic medical record
Insurance Type	Demographic survey	AYA self-report	Insurance type (e.g. private, public, or dual insurance)
AYA's Age	Demographic survey	AYA self-report	Age in years
Family Caregiver's Education	Demographic survey	Family caregiver self-report	Highest level of education obtained by each family member and noted on genogram
Parent's Marital Status	Genogram	AYA self-report	Type of marital status (e.g. married, divorced, separated, remarried, and single-parent)
<i>Independent Variables - (Process Variables)</i>			
		Description of Tools	Psychometric Properties
Self-efficacy	Sickle Cell Self-Efficacy Scale (SCSES)	9-item scale measuring an individual's perception of their ability to manage SCD on a Likert scale	<ul style="list-style-type: none"> Internal consistency (over all Cronbach's alpha = 0.89) (Edwards, Telfair, Cecil, & Lenoci, 2000) Divergent, construct, and predictive validity (Edwards et al., 2000)
Decision-making involvement	Decision-Making Involvement Scale (DMIS) (AYA-DMIS and Family caregiver-DMIS versions)	Two subscales (7-items in total) which assessed AYA seeking and expressing behaviors utilizing a Likert scale	<ul style="list-style-type: none"> Internal consistency (range of Cronbach's alpha from 0.71-0.91) (Miller & Harris, 2012) Temporal and construct validity (Miller & Harris, 2012)
<i>Dependent Variable - (Outcomes Variable)</i>			
		Description of Tool	Psychometric Properties
Transition readiness	Readiness to Transition Questionnaire (RTQ) (AYA-RTQ and Family Caregiver versions)	A tool with three subscales (1) measuring perception of transition readiness, (2) AYA healthcare responsibility levels, and (3) parent involvement utilizing a Likert scale	<ul style="list-style-type: none"> Internal consistency (range of Cronbach's alpha from 0.79-0.94) (Gilleland, Amaral, Mee, & Blount, 2012) Inter-rater reliability (Gilleland et al., 2012) Construct validity (Gilleland et al., 2012)

Table 10

Sample Characteristics

Variable	n(%) orM(SD)
AYA age	17.12 (SD 2.90)
AYA gender (female)	29 (58)
AYA SCD type	
HbSS (1)	40 (80)
HbSC (2)	7 (14)
HbS β + Thalassemia	3 (6)
HbS β 0 Thalassemia	0 (0)
Acute care visits in prior year	2.64 (SD 3.48)
Family caregiver gender (female)	44 (88%)
Family caregiver education level	
High school	13 (26)
Some college	13 (26)
College	14 (28)
Graduate school	10 (20)
Number of primary caregivers	
One family caregiver	43 (86%)
Two or more family caregivers	7 (14%)
Siblings provide support (Yes)	31 (62%)
Parents marital status	
Single parent	18 (36%)
Married	26 (52%)
Divorced/separated	3 (6%)
Remarried	3 (6%)
Insurance Type	
Private	22 (44%)
Public	27 (54%)
Dual	1 (2%)

Table 11

Correlation between AYA Decision-Making Involvement and Self-Efficacy

Variable	Self-Efficacy
DMIS: Child Express (AYA report)	0.481**
DMIS: Child Seek (AYA report)	0.208
DMIS: Child Express (Family caregiver report)	-0.052
DMIS: Child Seek (Family caregiver report)	0.161

** Correlation is significant at the 0.01 level.

Table 12

Significant Predictors of AYA Perceived Transition Readiness

Variables which significantly predicted AYA's self-reported transition readiness (n = 50).					
Dependent: AYA's self-reported TR (mean: 2.42 ± 0.70)	B	Standard error	Beta (R ²)	t	p
Gender (reference group: males)	-0.385	0.149	-0.276	-2.580	0.014
AYA age	-0.085	0.038	-0.356	-2.219	0.033
SCD type (reference: HbSS)	0.521	0.184	0.303	.2832	0.007
DMIS Subscales					
AYA reported child seek	0.275	0.113	0.374	0.2434	0.020
AYA reported child express	-0.409	0.148	-0.454	-2.757	0.009
AYA reported joint decision	-0.408	0.019	-0.388	-2.936	0.006
Self-efficacy	0.036	0.016	0.304	2.259	0.030
AYA self-reported healthcare responsibility	0.685	0.154	0.704	4.450	0.000
FC self-reported parent involvement	-0.503	0.146	-0.583	-3.443	0.001
Insurance type: (reference: private insurance)	-0.383	0.148	-0.276	-2.581	0.014

R² = 0.659, Adjusted R² = 0.548, F = 5.958, p < 0.000.

^a AYA: adolescent/young adult

Table 13

Predictors of AYA Self-Reported Healthcare Responsibility

Variables which significantly predicted AYA's self-reported healthcare responsibility (n = 50).					
Dependent: AYA's self-reported healthcare responsibility (mean: 2.69 ± 0.71)	B	Standard error	Beta (R^2)	t	p
AYA age	0.106	0.029	0.432	3.682	0.001
DMIS subscales					
AYA reported child express	0.287	0.101	0.310	2.835	0.007
AYA reported joint decision	0.255	0.103	0.236	2.481	0.017
Self-efficacy	-0.033	0.014	-0.273	-2.468	0.018
AYA reported parent involvement	-0.326	0.117	-0.317	-2.782	0.008
AYA self-reported TR	0.313	0.102	0.304	3.068	0.004
$R^2 = 0.674$, Adjusted $R^2 = 0.628$, $F = 14.815$, $p < 0.000$.					
^a AYA: adolescent/young adult					
^b TR: transition readiness					

Table 14

Predictors of Family Caregiver Perceived AYA Transition Readiness

Variables which significantly predicted family caregiver's perceived AYA transition readiness (n = 50).					
Dependent: FC perceived AYA's TR (mean: 2.17 ± 0.77)	B	Standard error	Beta (R^2)	t	p
DMIS FC reported child express	0.265	0.098	0.232	2.713	0.009
AYA self-reported TR	0.314	0.097	0.283	3.254	0.002
FC reported AYA healthcare responsibility	0.624	0.086	0.674	7.278	0.000
$R^2 = 0.750$, Adjusted $R^2 = 0.722$, $F = 26.392$, $p < 0.000$.					
^a AYA: adolescent/young adult					
^b TR: transition readiness					
^c FC: family caregiver					

Table 15

Predictors of Family Caregiver Perceived AYA Healthcare Responsibility

Variables which significantly predicted family caregiver's perceived AYA healthcare responsibility (n = 50).

Dependent: FC perceived AYA healthcare responsibility (mean: 2.38 ± 0.84)	B	Standard error	Beta (R ²)	t	p
SCD type (reference: HbSS)	-0.314	0.155	-0.152	-2.025	0.049
DMIS FC reported joint decision	0.274	0.102	0.193	2.694	0.010
FC reported parent involvement	-0.306	0.098	-0.295	-3.128	0.003
FC reported AYA TR	0.655	0.099	0.607	6.629	0.000

R² = 0.790, Adjusted R² = 0.755, F = 22.518, p < 0.000.

^a AYA: adolescent/young adult

^b TR: transition readiness

^c FC: family caregiver

^d SCD: sickle cell disease

CHAPTER SIX

CONCLUSION

Transition to adult healthcare has been linked with an alarming increase in morbidity and mortality for young adults living with sickle cell disease (SCD) (Blinder et al., 2015; Blinder et al., 2013; Brousseau, Owens, Mosso, Panepinto, & Steiner, 2010; Dickerson, Klima, Rhodes, & O'Brien, 2012; Lanzkron, Carroll, & Haywood, 2013; Paulukonis et al., 2014; Quinn, Rogers, McCavit, & Buchanan, 2010). Young adults with SCD post-transition have the highest rates of acute care and emergency room utilization related to SCD complications (Blinder et al., 2015; Dickerson et al., 2012; Quinn et al., 2010). In addition, there is a significant increase in mortality for young adults with SCD just after transition to adult healthcare (Blinder et al., 2013). With over 93% of youth with SCD living into adulthood, it is critical that action be taken to reduce poor health outcomes associated with transition to the adult healthcare setting (Quinn et al., 2010). Within clinical practice and research, efforts to understand and increase transition readiness in adolescents and young adults (AYA) with SCD have been undertaken to decrease bad health outcomes after transition. These efforts intend to ensure that AYAs have adequate knowledge and disease management skills to identify and engage with their future adult healthcare providers.

Development of transition readiness in AYAs has been endorsed as an essential milestone to reach prior to transfer to adult healthcare. Transition readiness is a measurement of the preparedness of the AYA to make the transfer to the adult healthcare setting. It encompasses having knowledge of their SCD, independently managing medication, learning to communicate with healthcare providers, and understanding how

to utilize healthcare resources. Transition is more than just a simple of transfer of care between pediatric and adult healthcare providers; it is a complex process which involves changing roles for multiple stakeholders including patients, family caregivers, and healthcare providers.

Even though transition has been recognized as a problem for almost three decades, interventions to improve transition readiness are not mature and are not being implemented consistently (Blum et al., 1993; Chu, Maslow, von Isenburg, & Chung, 2015; McManus et al., 2013; Zhou, Roberts, Dhaliwal, & Della, 2016). Research on transition has been conducted in a variety of settings and patient populations without the use of consistent methodological approaches including measures of transition readiness (Zhang, Ho, & Kennedy, 2014; Zhou et al., 2016). This challenges our ability to compare findings and generalize transition readiness research across populations and to move the science of transition forward.

Dissertation Overview and Findings

This dissertation study focused on examining the relationships between decision-making involvement, self-efficacy, and transition readiness in AYAs living with SCD prior to transition to adult healthcare settings. As noted in Chapter 2, there is limited information on modifiable factors associated with transition readiness hampering our ability to develop interventions to improve transition readiness prior to transfer to adult healthcare (Varty & Popejoy, 2020). Based on this analysis there was a clear need to identify additional factors related to transition readiness that could be targeted in transition interventions. To further understand what factors may be important to improving transition readiness, I carried out a metasynthesis of studies exploring

experiences and expectations of transition in AYAs living with a chronic disease (Chapter 3). The dominant theme in the metasynthesis was for better processes in transition preparation. Chapter 4 is the research proposal that explored the following aims: a) to explore the relationship between decision-making involvement and self-efficacy in AYAs with SCD, b) to explore the relationship between family caregiver's perception of decision-making involvement and self-efficacy for AYAs with SCD, and c) to predict transition readiness based upon AYAs' decision-making involvement and self-efficacy after controlling for AYA and family characteristics.

Approach

A descriptive, cross-sectional study was conducted with 50 family caregiver-AYA dyads. A total of 25 adolescents (ages 13-17) and 25 young adults (ages 18-21), and 50 family caregivers were recruited. Recruitment took place in a large urban comprehensive sickle cell clinic in Washington, District of Columbia between October 2019 and February 2020. The data was collected using the Decision-Making Involvement Scale, the Sickle Cell Self-Efficacy Scale, the Readiness to Transition Questionnaire, a demographics survey, and the electronic medical record. Data were analyzed in Statistical Package for Social Sciences Version 25 using Pearson product-moment correlations and multiple linear regression.

Major Findings

Data analysis demonstrated that there exists a positive relationship between AYAs reporting expressing opinions or ideas during decision-making and AYA self-efficacy regarding disease self-management ($r = 0.481, p < 0.000$). There were no significant relationships between AYA report of seeking information during decision-making, family

caregiver perception of AYA decision-making involvement (either expressing or seeking), and AYA self-efficacy regarding disease self-management. This study also established that there is a positive relationship between AYA decision-making involvement and AYA self-reported healthcare responsibility ($\beta = 0.310, p = 0.007$), and that there is a negative relationship between AYA reported parent involvement levels and AYA self-reported healthcare responsibility ($\beta = -0.317, p = 0.008$). In addition, family caregiver reported parent involvement level is negatively related to family caregiver perceived AYA healthcare responsibility level ($\beta = -0.295, p = 0.003$).

This study replicated findings established in prior studies including: a) a positive relationship between AYA self-efficacy and AYA expressing behaviors in decision-making involvement ($r = 0.481, p < 0.000$) (Miller, 2017), b) a positive relationship between AYA self-efficacy and AYA self-reported transition readiness ($\beta = 0.304, p = 0.030$) (Treadwell et al., 2016), c) a positive relationship between being female and transition readiness ($\beta = -0.276, p = 0.014$) (Treadwell et al., 2016), and d) a positive relationship between AYA age and AYA self-reported healthcare responsibility ($\beta = 0.106, p = 0.001$) (Speller-Brown et al., 2015).

There were a few unexpected findings. First is the inverse relationship between age and transition readiness. This finding may be an artifact of the setting, where standardized transition education starting at age 18. Potentially as AYAs received more information about transition and the challenges they faced with this change, the less ready they felt for the impending transfer. Having more knowledge may have increased their anxiety or concerns about the differences after transition to adult healthcare because they were fully aware of what they would be facing in the adult healthcare system. When

designing transition interventions, clinicians should consider adding an assessment of the AYAs feelings towards transition. Increased exposure to the changes expected in transition while increasing knowledge and skills essential for transition may also increase negative perceptions of the AYS's readiness to transition. Both health behaviors and perceptions of feeling ready should be assessed.

The second unexpected finding was the inverse relationship between self-efficacy and AYA self-reported healthcare responsibility levels. It was expected that higher self-efficacy would lead to greater healthcare responsibility but the inverse was found. As previously stated, there was a positive relationship between self-efficacy and AYA self-reported transition readiness, so to find that self-efficacy had the opposite relationship to the actual behaviors of AYAs is important to recognize. Self-efficacy had a positive impact on how they felt about transition but it did not demonstrate a positive impact on behaviors. This will be important to consider when designing future transition intervention which includes self-efficacy as a factor.

Discussion

The strengths of this study include the dyadic approach to data collection and the use of genograms to determine recruitment. With the genograms, this study was able to identify who was best to recruit within the family rather than relying on the traditional model of AYA-parent dyads. In addition, by including both the AYA and family caregiver, we were able to firmly establish that both AYA and family caregiver's perceptions of parent involvement were negatively associated with AYA healthcare responsibility. While perceptions between AYA and family caregivers varied for other factors, the results regarding parent involvement demonstrate that this is a critical

variable to address when preparing AYAs for transition to adult healthcare. These findings reaffirm the importance of starting transition preparation early as the process of shifting responsibility from family caregivers to an AYA will take time and frequent reinforcement. This process involves not only ensuring the AYA has sufficient knowledge and skills to carry out disease management, but it also involves teaching family caregivers to be mentors for their AYA.

This study also highlighted the possibility for interventions targeting AYA self-efficacy and decision-making involvement to improve their healthcare responsibility. By increasing the AYA's confidence in their role and encouraging them to express their ideas or opinions more frequently, it may increase their opportunity to learn how to become advocates for themselves. By taking active roles in decision-making about disease management, they are opening the door to more important conversations with their family caregivers about their need to develop more responsibility and independence. AYAs who are more confident may also engage more actively with their healthcare providers during visits. This could prepare them for the adult healthcare setting where adults are expected to speak for themselves.

Like parent involvement, AYA self-efficacy and decision-making is likely influenced by both the AYA and family caregiver. AYAs need to know that it is okay to express themselves more and should be encouraged to take more active roles in decision-making. They need to gain confidence in the practice of decision-making as adult healthcare providers will expect them to be independent in this process. At the same time it is important to recognize that family caregivers could view increased AYA self-efficacy or decision-making involvement negatively. They might be fearful of letting go

or still wish to be in control of disease management. Family caregivers should be prepared for this change, and interventions should encourage them to mentor their AYA and provide them with support as the AYA is taking on more responsibility. Interventions should help the family caregiver transition to their new role as a supportive caregiver and away from their old role as a primary caregiver.

This study had a few limitations. The recruitment setting's systematic transition education may have introduced bias into the AYA and family caregiver responses regarding transition readiness and healthcare responsibility. In addition, the use of self-report for measurement may have decreased the internal validity of the findings as AYAs and family caregivers may have under or over reported any of the study variables. Selection bias is also a threat to external validity as the study participants were recruited from a large, urban clinic with a transition education program, so the findings may not be generalizable to AYAs living with SCD in rural areas or those who receive care from clinics without transition education. Finally, this study was only able to sample participants who attended their scheduled appointments. The findings may not be generalizable to AYAs who frequently miss routine care.

Future Research

This dissertation study (Chapter 5) along with the systematic review (Chapter 2) and metasynthesis (Chapter 3) provided new insight and knowledge on the science of transition preparation, but it also highlighted a variety of gaps in our knowledge for AYAs living with SCD. Chapter 2 demonstrated the need to identify more modifiable factors which have associations with transition readiness. While the dissertation study (Chapter 5) found AYA self-efficacy, decision-making involvement, and parent

involvement to be promising variables for intervention development, the cross-sectional design of the study does not provide any insight into how these factors affect transition readiness over time. Longitudinal studies are needed to assess how modifiable factors and transition readiness influence health outcomes over time and post-transition. Emphasis has been placed on understanding and nurturing transition readiness in AYAs prior to transition; however, studies have not established that transition readiness predicts positive health outcomes. To best development transition interventions, it is critical that factors which could increase positive health outcomes post-transition be identified for future interventions.

There is a gap in health outcomes research regarding AYAs with SCD post-transition. Chapter 2 showed that little is known about how variables associated with transition readiness, or transition readiness itself, relate to positive health outcomes. If efforts are going to be taken to prepare AYAs living with SCD for transition, these efforts need to be evidence based and chosen for their ability to improve health outcomes post-transition. While Chapter 5 established potential modifiable factors for transition interventions, the ability of these factors to support successful transition is still unknown. Healthcare providers and researchers can expand upon the current body of transition literature by incorporating both pediatric and adult healthcare providers into future research. A majority of the literature on transition was developed by pediatric providers, but future research needs to explore transition through the lens of both pediatric and adult healthcare providers. There needs to be discussion of how to create seamless care between the two settings and which variables are important to both settings. Research on adult provider's expectations of young adults may shape how we prepare AYAs for

transition and could assist with identifying ways to improve adult healthcare approaches to engaging with and supporting young adults living with SCD after transition.

One final takeaway from the collective work within this dissertation is that transition readiness has different definitions to different stakeholders. To healthcare providers, transition readiness focuses on the AYA's level of disease knowledge and self-management skills at a specific time. It does not take in account the AYAs' feelings of unpreparedness. Within the metasynthesis (Chapter 3), AYAs described variety of emotional barriers to being ready to transition. AYAs reported feelings of grief, anxiety, sadness, and a lack of necessary knowledge and skills to succeed after transition (Bemrich-Stolz, Halanych, Howard, Hilliard, & Lebensburger, 2015; Porter, Graff, Lopez, & Hankins, 2014; Varty, Speller-Brown, Phillips, & Kelly, In review). While the definition of transition readiness used within healthcare is critical as this knowledge and these skills will be expected of them by their healthcare providers, a more comprehensive definition should be considered. AYAs ought to be prepared with the right information, skills, and emotional support so they can successfully transition and develop strong relationships with their new adult healthcare providers just as they had accomplished with their pediatric providers.

In conclusion this dissertation provided new knowledge of modifiable factors which should be considered when developing interventions to improve transition readiness in AYAs living with SCD. Though there are still many gaps to be explored, these findings are promising. With further research and the implementation of evidence-based intervention to increase transition readiness, transition in the future could become a

coordinated, patient-centered process which results in positive health outcomes for all AYAs living with SCD.

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VITA

Maureen M. Varty is a native of St. Louis, Missouri and was raised by a mother who is a doctoral prepared nurse and her father who is a physician. She has two sisters who are also in the nursing profession. She received her Bachelor of Science in Nursing from Saint Louis University in 2014, and she has over five years of clinical experience in acute care nursing. She has been actively involved and assumed leadership roles in hospital-wide nursing led committees focused on healthcare legislation and pain management of the acute care patient population. In addition, she has served as a policy advocate for the Epilepsy Foundation of Missouri and Kansas, the Nursing Community Coalition, and the American Association of Colleges of Nursing affording her the opportunity to advocate for policy changes on the state and federal level. Through both her personal health experiences and her clinical experiences of poor transitions for youth living with chronic diseases she decided to pursue her doctoral degree in 2016 at University of Missouri-Columbia Sinclair School of Nursing. During her doctoral program she successfully has her dissertation proposal funded by the National Institutes of Nursing Research. She currently resides in Westminster, Colorado with her husband who is an IT analyst for the Government Accountability Office. Maureen works as a nurse scientist at UCHealth University of Colorado Hospital in Aurora, Colorado.