YOUNG ADULTS WITH SICKLE CELL DISEASE IN PEDIATRIC CARE: CRISIS
IN TRANSITIONING TO ADULT CARE

by

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and approved by

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ABSTRACT

YOUNG ADULTS WITH SICKLE CELL DISEASE IN PEDIATRIC CARE: CRISIS IN TRANSITIONING TO ADULT CARE

By PAULETTE E. FORBES

Dissertation Director:
Sabrina M. Chase, PhD

Sickle cell disease (SCD) is the most common genetic disorder in the United States, primarily affecting individuals of African descent. It is characterized by anemia and a constellation of acute and chronic complications that result in increased morbidity and mortality. Recent advances in medical technology, diagnosis and medical management have resulted in extending life expectancy from the early teens into the fourth and fifth decades. As a result, although SCD was once considered a health condition of childhood, it now poses challenges for young adults who have survived the illness and must now transition from the pediatric to the adult health care system.

This mixed-methods study explored the transition experiences of young adults with SCD from the perspectives of both young SCD patients and health care providers. It was guided by the following research questions: (1) What are the individual, provider and system-level factors that young SCD patients perceive as facilitating and/or impeding transition from the pediatric to the adult health care system? (2) Are young adults with SCD being prepared to access and navigate the adult health care system? (3) What are health care providers’ perceptions of the patient, provider and system-level factors that
impact transition? (4) How and when should transition from pediatric to adult health care services begin?

The study included both qualitative and quantitative components. Sixteen young adults with SCD ranging from 18 to 30 years of age were interviewed using a semi-structured interview guide. Content analysis was utilized to generate codes and identify themes. Additionally, a 16-question web-based survey was administered to 38 health care providers (including nurses, physicians and nurse practitioners) from the pediatric hematology, adult hematology and pediatric and adult emergency departments.

Young SCD patients were very satisfied with the care they received in the pediatric health care system. During interviews, they described developing trusting relationships with pediatric providers and non-clinical team members who were easily accessed during emergencies of all kinds. They also reported fear of leaving pediatrics, minimal or no preparation for the adult health care system, long waits for adult hematology appointments and being stereotyped as drug seekers during emergency department visits. Although they were satisfied with their health care provider(s) in the adult clinic, their transition to the adult health care system was characterized by a reduced access to regular care as a result of limited clinician office hours, chronic specialist understaffing and the absence of ancillary or support staff. All patient participants reported long time wait times for evaluation and treatment in adult emergency rooms coupled with inadequate pain management. Health care providers who completed the survey identified the same barriers as described by patient participants. Providers acknowledged “excellent clinical care” in the pediatric system and a cluster of barriers in the adult system. These included a lack of adult providers and SCD specialists, failure to
prepare young adults for transition and system-level barriers such incompatible electronic medical record platforms that inhibit information sharing. These findings underscore the necessity of implementing a comprehensive multidisciplinary transition program to prevent increased morbidity and mortality, decrease emergency department use and improve the quality of life for young adults with SCD who age out of pediatric care.
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DEDICATION

This dissertation is dedicated to all the young adults with sickle cell disease who have aged-out of pediatric care, especially those who may be lost on their journey to adult care.
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Sickle cell disease (SCD) is the most common genetic disorder in the United States. It is the first disease for which a genetic mutation was identified and the first molecular disease that was described (Wailoo, 1997; Pauling et al., 1949). Approximately 70,000 to 100,000 persons in the United States have the disease (Heeney & Ware, 2008). Although SCD is seen in individuals of Mediterranean, Middle Eastern, Indian, Caribbean and Hispanic ancestry, it is found primarily in persons of African descent. In the United States, ninety-five percent of all cases occur in people identified as black or African American. Approximately nine percent of the African American population are carriers of the gene (Heeney & Ware, 2008; National Heart, Lung and Blood Institute, 2002; Platt et al., 2002). An estimated 2,000 infants are identified annually with SCD in the US (Buchanan et al., 2010).

Approximately one in 12 African Americans and about one in 100 Hispanic Americans carry the sickle cell trait, which means they are carriers of the disease. When two individuals who are carriers of the defective gene have a child, the child has a 25 percent chance of having the disease and a 50 percent chance of being a carrier. The disease occurs in about 1 of every 500 African American births, and about 1 in every 1000 to 1400 Hispanic births in the United States (Mann-Jiles & Morris, 2009).

Sickle cell disease (SCD) is a chronic condition with a wide range of symptoms and complications. Symptoms include acute pain (vaso-occlusive crisis), chronic pain, pulmonary, renal, cardiac, ophthalmological and neurologic issues. Complications can
include iron overload, bone infarction and avascular necrosis. In 1973, the average life span of an individual with SCD was 14 years of age. Today, life expectancy extends into the fifth and sixth decades (Claster & Vichinsky, 2003; Platt et al., 1994). This change in mortality and morbidity is due, in part, to early detection through newborn screening and other medical innovations such as prophylactic antibiotic therapy, improvements in self-care, and transfusion therapy (Powars et al., 2005; Davis et al., 1997).

Despite these advances, the survival of young adults with SCD is associated with a number of health problems and adverse health outcomes. These include the predisposition to life threatening infections, cardiomyopathy and cardiac failure, pulmonary hypertension, chronic lung disease, high morbidity in pregnancy, stroke and kidney failure (Fixler & Styles, 2002; Steinberg, 1999). Individuals with SCD are at increased risk of experiencing several of these complications during their lifetime. Therefore, the delay or lapse in health care services as one enters adulthood is detrimental to survival. For example, individuals with SCD (especially those with Hgb SS) are at high risk of severe bacterial infections because the spleen, which is usually afunctional by the age of 10 years or earlier as a result of splenic infarctions, is not able to fight infections with encapsulated bacteria like *Streptococcus pneumoniae* (Klings et al., 2015). Associated conditions like pulmonary hypertension, cardiomyopathy and left ventricular dysfunction are major causes of sudden cardiac death. A study done by Darbari and colleagues that examined circumstances of death over a twenty-five-year period found that 23.4% of adult deaths from SCD was sudden (James et al., 1994; Darbari et al., 2006).
Creating comprehensive transition pathways for young adults with SCD to ensure that they continue to receive optimal health care services is critical. This is particularly important because young adults are at highest risk for early mortality (Quinn et al., 2010). Continuity of care is therefore essential for improving patient outcomes for this population of patients. Patients with SCD between the ages of 18 to 30 years have higher readmission hospital rates, up to 48.3% more than other individuals in same age group (Brousseau et al., 2010b). These findings support the assertion that moving from pediatric to adult health care services is often unplanned and poorly organized. As a result, individuals are at risk for receiving suboptimal care or receiving no care at all and thereby experiencing negative effects on morbidity and mortality.

Health care transition was defined more than 20 years ago as “the purposeful planned movement of adolescents and young adults with chronic medical and physical conditions from child-centered to adult-centered health care services” (Blum et al., 1993). As more young people with chronic illnesses survive into adulthood, the issue of transition poses new challenges to the healthcare system, which has led to numerous recommendations. In 2002, the American Academy of Pediatrics (AAP), the American Academy of Family Physicians (AAFP), and the American College of Physicians – American Society of Internal Medicine developed a consensus statement on healthcare transition for adolescents and young adults with special health care needs:

“The goal of this policy statement was to ensure that by the year 2010 all physicians who provide primary care or subspecialty care to young people with special health care needs understand the rationale for transition from child-oriented to adult-centered care; have the knowledge and skill to facilitate that process; and know if, how and when transfer of care is indicated” (American Academy of Pediatrics [AAP], 2002b, pp. 1304).
However, more than a decade after the target date for achieving that goal, the health care system continues to struggle with how to create effective pathways for young people with chronic illnesses to transition from pediatric to adult health care services.

The process of moving from the pediatric to the adult health care system is an important and necessary step to delay the morbidity and mortality associated with chronic illnesses. The development of comprehensive, multidisciplinary transitional care models that promote seamless and effective transfer of medical care from the pediatric to the adult health care system are urgently needed. Such models of care will ensure that access to continued health care services is uninterrupted as young adults move from pediatric to adult care.

1.2. The Research Problem

In 1993, Blum and colleagues argued that the goal of transition planning is to provide comprehensive, continuous health care that is developmentally appropriate and meets the psychosocial and family needs of adolescents and young adults with pediatric onset diseases. Despite recommendations and policies, the American health care system is still unable to develop and deliver successful, comprehensive transition services for young people with pediatric onset chronic conditions like SCD. Most children with SCD are treated in comprehensive pediatric sickle cell disease centers. These centers lack the structure and guidance of a well thought out transition model that can provide information and guidance to adolescents and young adults on how to best navigate the adult health care system, especially in an environment of health insurance constraints, health care specialists’ unavailability, and health care cost containment.
Although some young adults are able to navigate the adult health care system, there is a significant proportion that experience difficulties with accessing and navigating the adult system. The consequences of ineffective transition range from a lack of continuity of care to receiving episodic, emergency care or dropping out of care completely. These consequences increase morbidity and mortality. Even institutions that have established transition programs are oftentimes unable to meet the goal of collaboration and coordination of services (Scal et al., 1999; Scal & Ireland, 2005). Consequently, patients too often either experience simple transfer to adult health care services or simply dropout of care (Viner, 1999). Research shows that young adults with chronic illnesses who do not have transition support from pediatric to adult health care services oftentimes experience poorer health, receive compromised quality of care and incur higher medical costs (Prior et al., 2014). Simple transfer of care, episodic care or not receiving medical care are approaches that do not conform to acceptable standards of care. Therefore, successful transition programs must meet the needs of young adults with pediatric onset chronic conditions like sickle cell disease, as well as health care providers in both the pediatric and adult departments who provide care to this population.

Adolescence is a time when individuals undergo significant psychological, physical and social developmental changes, and those with chronic illnesses have the added burden of making the transition to the adult health care setting. Moving from child-centered health care (where care is family-centered) to adult health care (where care is individualized) poses one of the greatest challenges in transitioning from pediatric to adult healthcare services (Hagood et al., 2005; Rosen, 2004). Several studies have demonstrated that transitioning from pediatric to adult health care services is oftentimes
haphazard and fragmented. Even self-identified transition programs can fail to achieve
the goal of comprehensive, collaborative transition (Scal et al., 1999; Wojciechowski et
al., 2002).

Although there are many consensus statements focusing on transition, there is a
lack of evidence-based guidelines concerning the best practices for successful transition
from pediatric to adult health care system for young adults with sickle cell disease.
However, studies that have examined patient satisfaction after transition have shown
these to be associated with better patient outcomes and quality of life (Sawyer, 1998;
Rettig & Athreya, 1991; Nasr et al., 1992). Furthermore, coordinated, uninterrupted
transfer of medical care from pediatric to adult health care services for young adults with
chronic conditions like SCD may have a beneficial effect on health care costs (Brotzman
et al., 2001). Prevention delivers real value as a cost-effective way to keep individuals
healthy and improve their quality of life. The prevention of disease complications will
also result in the prevention of disability and premature death (Viner, 2008).

1.3. Purpose and Significance

The objective of this study is to describe individual, provider and facility
characteristics that promote or hinder health care transition from pediatric to adult
healthcare services for young adults with sickle cell disease in one tertiary care institution
in central New Jersey. The study explores how young adults aged 18 to 30 years,
experience movements from the pediatric to the adult health care system. It also
describes the individual, provider and system factors that health care providers perceive
to be the factors that hinder and/or promote transition from pediatric to adult health care system.

Since pain is the hallmark of SCD, the emergency department is oftentimes the first point of contact when an individual with SCD experiences any complication of the disease. To this end, getting the perception of emergency room clinicians is important to include in this study. The initial proposal included young adults with SCD who were seen in the emergency department but had no regular source of SCD health care services. However, recruitment of the latter population was unsuccessful. Studies have focused on patients’ perceptions and health care providers’ perception of transition, but none have focused both on patients’ and providers’ perceptions of health care transition from pediatric to adult services in a single tertiary institution that has the infrastructure to provide health care services to both pediatric and adult populations (Anderson et al., 2002; Scal, 2002). Having both pediatric and adult health care providers within the same health care system, located on the same campus and who have sickle cell disease expertise are important elements that are necessary to ensure seamless health care transition for this population of patients. The goal of this study is to fill this gap by using qualitative and quantitative techniques to identify and understand the patient, health care provider and health care facility factors that promote and/or hinder effective transition from pediatric to adult health care service for young adults with sickle cell disease.

Researchers have examined transition from the perspectives of children, adults, caregivers and health care providers but has examined them as separate populations (Telfair et al., 2004; Scal, 2002; Scal & Ireland, 2005; Hauser & Dorn, 1999; Patterson & Lanier, 1999). By investigating the perspectives of young adults aged 18-30 years with
SCD in both the pediatric and adult systems, and the health care providers who care for them within the same health care system, this research elucidated the factors that facilitated and impeded seamless transition of care from the pediatric to the adult health care system. An examination of the problem in a single tertiary care facility that has both pediatric and adult hematologists (physicians who specialize in the treatment of blood disorders and malignancies) provided critical insights into what patients and providers perceive as the facilitators and barriers to effective transition for young adults with sickle cell disease.

The Healthy People’s 2020 goal of transitioning, reducing and ultimately eliminating health disparities experienced by minorities has relevance to this study. Health disparities among Blacks and other racial groups exist in life expectancy, death rates and other measures of health status. In 2010, the percentage of Black adults living in poverty was among the largest as compared to other ethnic groups (Agency for Healthcare Research and Quality, 2012). Factors such as poverty, education level, insurance status, the availability of transportation, health care providers who have experience caring for individuals with SCD, and clinical hours of operation are some of the factors that contribute to health disparities, especially in a population that already is at risk for poorer health outcomes.

Although SCD affects individuals of other ethnic groups, the burden of the disease is largely borne by Blacks and individuals of Hispanic origin. These two groups are also at increased risk for experiencing disparities in health care services as is evidenced by higher rates of disease and deaths than their White counterparts (Williams & Sternthal, 2010). It is also well documented that differences in income and education
contribute to health disparities, with those with lower income and less education experiencing more negative outcomes (Williams et al., 2010). Consequently, some of these factors are explored with the patient population during the course of the study.

1.4. Research Questions

The central questions addressed by this study are:

1. What are the individual, provider and system factors that patients perceive as facilitating and/or impeding transition from pediatric to adult health care system for young adults with SCD?

2. Are young adults with SCD being prepared to access and navigate the adult health care system?

3. What are health care providers’ perceptions of patient, system and providers’ factors that impact transition?

4. How and when should transition from pediatric to adult health care services begin?

The research also attempted to answer the following sub-questions as they relate to young adults with sickle cell disease and health care providers who provide SCD-related care to them:

a. How are adolescents prepared to access healthcare services in the adult system?

b. How knowledgeable are the patients with SCD about their disease and do they understand their treatment plan?

c. How do patients experience transition?
d. What are the perceived reasons concerning why adults remain in pediatric care? (individual and provider barriers).

e. What are the perceived system barriers expressed by patients and clinicians?

f. How did the young adults who are in adult care get there and what if, anything, they would do differently?

g. What recommendations would they propose (clinicians and patients) to improve transition?

1.5. Structure of Dissertation

This dissertation describes data that provide answers to the research questions and proposes a best practice template for transitioning from pediatric to adult health services for young adults with sickle cell disease. Chapter Two provides a detailed review of the sickle cell literature that includes pathophysiology and complications of sickle cell disease, and the importance of ensuring that health care services are uninterrupted when young adults age out of pediatric care. It also outlines the organization of pediatric sickle cell disease care in comprehensive sickle cell centers in New Jersey and provides a detailed description of the current state of SCD care at one tertiary care facility in central New Jersey. The chapter also provides a detailed review of the health care transition literature, which includes the benefits of effective transition, the negative health consequences of unsuccessful transition, barriers to transition and models of transition that have been used for children and young adults with pediatric onset chronic illnesses like SCD. A description of the theoretical underpinning of the research project is discussed. Chapter Three outlines the study design, study setting, development of the
research tools, procedures for data collection and the treatment of the data. Chapters Four and Five explain the procedures for analysis of the data obtained from patients and health care providers and discusses the findings. Chapter Six merges the findings of patients and health care providers and integrates the findings of the study to provide a strong argument for the reasons why young adults are still receiving care in pediatrics. It also summarizes the research findings including the strengths, weaknesses, the limitations of the study.

As part of data collection, responses were elicited from both patients and providers regarding what would work to resolve the problem of transitioning young adults with SCD. The *BRIDGE Model of Care* was developed based on the findings of the study. There is a concise description of the proposed model and an outline of how the model could be implemented in a tertiary care facility that has pediatric and adult hematologists on the same campus, as well as having the health care infrastructure to facilitate such a model. Healthcare infrastructure is defined as the physical facilities, trained staff, and the capacity to provide preventive care, health surveillance and health promotion, as well as treat acute and chronic health conditions. The proposed model, if piloted, has the potential to assist in the resolution of the problem. The researcher also makes suggestions for future studies.
CHAPTER TWO

REVIEW OF LITERATURE

This literature review examines the transition of adolescents and young adults from pediatric to adult health care system. It also focuses on SCD and the complications that confer the most morbidity and mortality on those who are afflicted. It discusses the provision of sickle cell disease medical care for the pediatric population and its organization in comprehensive sickle cell centers. It also describes the current state of SCD health care services within one tertiary care facility in central New Jersey.

2.1. Sickle cell disease

Dr. James Herrick first described sickle cell disease in the medical literature in 1910 (Herrick, 1910). The disease was characterized at the molecular level in the 1950s (Ingram, 1957). It was the first disease for which a molecular defect in a gene was identified and it is the most common genetic disease diagnosed as part of the U.S. Newborn Screening Program (Therrell, 2015). Forty-four states, the District of Columbia, Puerto Rico and the Virgin Islands provide universal newborn screening for SCD; screening is available by request in the other six states (AAP, 2002a). There are approximately 2.5 million individuals in the United States who are heterozygous carriers of the sickle cell gene. The prevalence of SCD surpasses that of all other serious genetic disorders (AAP, 2002a).

SCD is an inherited disorder that involves genetic malformation of the red blood cells. Normal hemoglobin is a flat, oval shaped cell that easily passes through the blood vessels. Sickle cells possess a defect in the gene that produces hemoglobin, the
component of the red cells that is responsible for carrying oxygen to the tissues and organs of the body. The abnormal hemoglobin within the red cells causes them to elongate and stretch into rod-shaped cells that are sticky and rigid, thereby impairing their ability to pass through the blood vessels. The sickled cells aggregate and clog the blood vessels, interfering with the delivery of oxygen to organs. This process results in ischemic injury to the tissue and sometimes tissue death, which is known as infarction or necrosis (Lisak, 1992).

This genetic hemoglobin disorder is caused by the inheritance of an altered beta-hemoglobin chain gene from both parents. When two individuals who are carriers of the defective gene have a child, the child has a 25 percent chance of having the disease and a 50 percent chance of being a carrier. Normal adult hemoglobin, called hemoglobin A, consists of four globin chains: two (alpha) \( \alpha \) globin chains and two (beta) \( \beta \) globin chains. Sickle cell anemia results from a mutation in the \( \beta \) globin gene (Platt, 2008). The inheritance of two sickle \( \beta \) globin genes, or one sickle \( \beta \) gene in combination with another \( \beta \) globin chain defect, results in SCD (Fixler & Styles, 2002). Individuals who inherit a single gene from one parent have a mixture of the Hgb S and normal hemoglobin Hgb A. Individuals with this combination have “sickle cell trait.” They are carriers of the disease, but they do not experience symptoms and complications of the disease like those who carry both defective genes and have sickle cell disease.

Normal Hemoglobin A (HgbA) contains glutamic acid in the sixth amino acid position of the beta chain of hemoglobin. Hemoglobin S (Hgb S) results from the substitution of a valine for a glutamic acid, and Hemoglobin C (Hgb C) results from the substitution of lysine for glutamic acid at the sixth position. Sickle cell disease denotes
all genotypes that have at least one sickle gene in which Hgb S makes up at least half of the hemoglobin that is present (Stuart & Nagel, 2004). There are many variants of SCD, which are combinations of Hgb S with $\beta^0$ or $\beta^+$ thalassemia, also known as sickle thalassemia (Bunn, 1997; Centers for Disease Control and Prevention, 1998; Dang et al., 2005; Stuart and Nagel, 2004). Although many sickle cell genotypes are linked to the disease, Hgb SS, Hgb SC and sickle beta thalassemia (S$\beta$ thal) are the most severe (Bunn, 1997; Centers for Disease Control and Prevention, 1998). However, Hgb SS is the most common variant and has the most severe clinical manifestations of the group. It accounts for 60%-70% of sickle cell disease in the US (Stuart and Nagel, 2004).

The lifespan of normal red blood cells (RBCs) is about 120 days, while that of a person with sickle cell disease is about 12 to 15 days (Platt et al., 2002). As a result, hemolysis, anemia and vaso-occlusion are clinical hallmarks of the disease (Wanko & Telen, 2005; Ballas, 2005).

Clinical symptoms of the sickle cell disease can range from mild to life threatening. Hemolytic anemia results from the premature destruction of the RBCs and the bone marrow’s inability to compensate for the destruction of these cells. As stated before, normal RBCs are flat and smooth; however, the crescent shape of sickle cells prevents them from acquiring adequate oxygenation. The deoxygenated hemoglobin S molecule causes the red cell to become rigid and entangled, thus causing blockage in the small blood vessels. The damaged RBCs also increase the viscosity of blood and this increased viscosity results in vaso-occlusion. Vaso-occlusion limits the flow of blood and the delivery of oxygen to vital organs and this results in ischemic injury. The resultant obstruction, ischemia, infarction and necrosis caused by sickled cells affects all organs of
the body and contributes to the major complications of the disease (Platt et al., 2002; Swerdlow, 2006; Fixler & Styles, 2002). Although vaso-occlusion is the hallmark of sickle cell disease, anemia caused by hemolysis (premature destruction of red cells), and increased inflammation also contributes to complications. These complications include pulmonary dysfunction, life-threatening infections, kidney dysfunction, neurological disorders, vascular, hepatic, bone and joint problems. These organ system complications oftentimes result in lifelong disabilities and even early death (Butler & Beltran, 1993; Lisak, 1992).

Advances in medical treatment and scientific technologies have made tremendous contributions in extending the lifespan of children who are born with chronic illnesses like sickle cell disease. In 1994, Platt et al. reported on a prospective analysis of deaths that occurred among patients in the Cooperative Study of Sickle Cell Disease (CSSCD) and found that the median age for survival for people with Hgb SS was 42 years for men and 48 years for women. The median age of survival for those with Hgb SC was even longer - sixty years for men and 68 years for women. Overall, fifty percent of patients with SCD survived into the fifth decade of life (Platt et al., 1994). There are also published reports of individuals with SCD who have survived into the seventh decade of life (Steinberg et al., 1995). Platt and colleagues (1994) also reported that adult patients with SCD who experienced frequent pain episodes tended to die earlier than those who do not.

1 CSSCD is a prospective study of the clinical course of SCD in which more than 3764 patients were enrolled from birth to 66 years of age at 23 clinical centers between September 1978 and 1988 in the US. It was sponsored by the National Institutes of Health (Platt et al., 1994)
From the 1980s, advances in the treatment of sickle cell disease was limited to newborn screening (Vichinsky et al., 1988), prophylactic use of the antibiotic penicillin (Gaston et al., 1986) and the use of hydroxyurea to ameliorate of some of the symptoms of sickle cell disease (Heeney & Ware, 2008). However, in 2017, the American Society of Hematology (ASH) introduced its advocacy initiative targeted at improving outcomes for individuals with sickle cell disease worldwide. Since that time, legislation aimed at leveraging increased funding for research, treatment and education has led to significant advances for a condition that lagged behind for over a century (American Society of Hematology, 2017). Some of these advances will be discussed later in this chapter.

2.2. Complications of Sickle Cell Disease

The complications of sickle cell disease are many and varied. They include pain; increased risk of life-threatening infections; pulmonary, cardiac, neurological and vascular dysfunction; life-threatening anemia, and bone and joint problems. These organ system complications can result in lifelong disabilities and death (Butler & Beltran, 1993; Lisak, 1992). As discussed earlier, the process of vaso-occlusion interrupts the supply of oxygen to many organs in the body and this in turn causes damage to the organs and surrounding tissue. Some of the complications can also be a result of treatment: for example, iron deposit in cardiac muscle and the liver as a result of chronic transfusion therapy.
2.2.1. Pain

Pain is the number one cause of hospitalizations for individuals with sickle cell disease, especially for adults (Ballas & Lusardi, 2005). It is the primary reason for seeking emergency medical care. An exacerbation of pain is referred to as “vaso-occlusive crisis.” Crises can vary in intensity, ranging from mild episodes to extensive, debilitating episodes that require prolonged hospitalization and the use of intravenous narcotics and rehydration fluids. These unpredictable and recurrent episodes of pain can occur in any part of the body. However, the areas that are most affected by sickle cell crisis are the long bones, ribs, sternum, spine and pelvis (Stuart and Nagel, 2004). Multiple sites can be affected at the same time.

Pain is initiated by obstruction of the small blood vessels with sickled red blood cells. The deformed red blood cells also increase the viscosity of the blood, thus retarding blood flow. This stasis promotes further sickling, which in turn results in tissue inflammation, ischemia, necrosis and infarction (Mehta et al., 2006; Platt et al., 2002). Pain by itself is debilitating, but the onset of a painful crisis can also be the catalyst for triggering the onset of complications in other organ systems such as the lungs. Pain episodes can be recurrent, unpredictable and vary in frequency, intensity, severity and duration. They can last days to weeks and there are individuals who have daily chronic pain. Although pain can occur without precipitating events, it is often triggered by exposure to extreme temperature (heat or cold), infection, fever and dehydration.
2.2.2. **Pulmonary Complications**

The lung is a major organ for complications of sickle cell disease. Pulmonary complications are among the most common causes of hospitalization for individuals with sickle cell disease. Complications can be both acute and chronic and are the leading cause of morbidity and mortality in the adult population (Castro et al., 1994; Vichinsky et al., 1997). Clinical lung involvement manifests in two major forms: acute chest syndrome (ACS) and pulmonary hypertension (PH). These occur as a result of infarction of the pulmonary parenchyma caused by blockage and hypoxia, fat embolism from bone marrow of infarcted bone, hemolysis and infection (Vichinsky & Styles, 1996; Scully, Mark et al, 1997). More than 20 percent of adults with sickle cell disease die as a result of pulmonary complications of the disease (Platt et al., 1994).

2.2.3. **Acute Chest Syndrome**

Acute chest syndrome (ACS) is the most common cause of prolonged hospitalization and is the leading cause of death for individuals with sickle cell disease. The hallmark clinical symptoms of ACS are chest pain, fever, cough and new infiltrate on chest radiograph (Vichinsky et al, 2000; Castro et al., 1994). The Cooperative Study of Sickle Cell Disease identified ACS as a leading cause of premature death among individuals with SCD. Acute chest syndrome is estimated to occur in 15 to 43 percent of individuals with SCD and recurs in as much as 80 percent of those who have had a prior episode. It is responsible for the deaths of as many as 25 percent of individuals with SCD and is the leading cause of death for individuals over 10 years old. Acute chest syndrome is a predictor of shortened lifespan and repeated episodes increase the risk of chronic lung
disease and pulmonary hypertension (Castro, et al., 1994; Platt et al., 1994; Powars et al., 2005).

2.2.4. Pulmonary Hypertension

Pulmonary hypertension is one of the serious complications of sickle cell disease. It is characterized by sustained elevated pulmonary artery pressure of 25 mmHg or greater (Shah, 2012). Persistent elevated pulmonary artery pressure ultimately results in heart failure, thus increased mortality.

The precise prevalence of pulmonary hypertension is unknown. However, prospective and retrospective studies strongly support the thesis that pulmonary hypertension is the greatest risk factor for early mortality for patients with SCD (Ataga et al., 2004; Gladwin et al., 2004; Castro et al., 2003; Sutton et al., 1994). Sutton and colleagues reported a 40 percent mortality rate at 22 months after diagnosis. Castro and colleagues (2003) confirmed this with similar findings; they reported a 50 percent mortality rate at 24 months. The poor prognosis that this complication confers highlights the importance of ensuring seamless transfer of medical care from pediatric to adult settings. Close surveillance of individuals with SCD is imperative because preventive measures can be implemented. Early recognition of symptoms is also essential to decrease morbidity and mortality and increase quality of life for these individuals.

2.2.5. Neurological Complications

The brain is a major organ that is affected by SCD. Strokes occur in individuals with all sickle cell genotypes. Cerebrovascular accident (CVA) or stroke is the leading
cause of disability and death for children and adults with SCD (Prengler et al., 2002; Ohene-Frempong et al., 1998; Platt et al., 1994). Cerebrovascular accident is the interruption of the blood or oxygen supply to the brain that results in neurological signs and symptoms. Occlusion of blood vessels (infarction) or rupture of a weakened blood vessel (aneurysm) causing hemorrhage is the pathophysiology of strokes (Hillery & Panepinto, 2004). It is estimated that by the age of 20 years, 11 percent of individuals with SCD have had a clinically apparent stroke and that risk increases to 24 percent by the age of 45 years old (Ohene-Frempong et al., 1998). Young adults (20-29 years) are most at risk for hemorrhagic strokes, which has a mortality rate of 26 percent in the first two weeks following the stroke (Wanko & Telen, 2005). The resultant neurological and cognitive impairments following a stroke can have significant negative impact on the patient, his/her family and health care costs.

Clinically overt strokes may impair cognitive and motor function. However, as many as 22 percent of children with Hgb SS have subclinical strokes, that is lesions in the brain that are detected on surveillance magnetic resonance imaging (MRI) or computed tomography (CT) scan. Children with ‘silent infarcts’ have decreased scores on neuropsychological tests that assess verbal, mathematical, and visual motor skills (Kinney et al., 1999; Adams, 2001). Cognitive deficits often have a negative impact on educational attainment and limit employment opportunities. Additionally, diminished cognitive functioning puts individuals with ‘silent infarcts’ at risk for experiencing greater difficulty in maneuvering the pathway from pediatric to the adult healthcare system. These individuals require appropriate preparation and guidance on how to
navigate the adult health care system and having access to a structured transition program is the best way to address this issue.

The current treatment for pediatric patients who have had neurological insult as a result of a stroke is chronic red blood cell transfusion therapy. This is done either by simple red blood cell transfusion or exchange transfusion. Exchange transfusion is the removal of the patient’s red blood cells and replacement with donor red cells. The goals of transfusion are to improve the blood’s oxygen-carrying capacity by correcting anemia and to prevent or reverse complications of SCD. For these individuals, chronic transfusion therapy is used to keep sickle hemoglobin levels below 30 percent because at this level there is a reduced the risk of recurrent stroke (Davis et al., 2017). It is estimated that 44 to 67 percent of individuals will experience recurrent infarctions after the first stroke (Balkaran et al., 1992).

The Stroke Prevention Trial in Sickle Cell Anemia (STOP) was a multi-center randomized clinical trial that was initiated in 1995 to examine if prophylactic blood transfusion could prevent strokes in children. It was terminated in 1997, which was earlier than anticipated because the results were so compelling. As a result of that study, the National Heart, Lung and Blood Institute (NHLBI) issued a clinical alert for the use of transcranial Doppler screening for all children aged 2 to 16 years because the study revealed that regular red blood cell transfusions reduced the risk of first stroke by 90 percent (Adams et al., 1998). Transcranial doppler uses ultrasound to detect the blood flow velocity in the intracranial cerebral artery (ICA) and the middle cerebral artery (MCA) for pediatric patients with SCD. An increase in the velocity is associated with increased risk of stroke (Purkayastha & Sorond, 2012).
The follow-up study, STOP–II trial, designed to determine if blood transfusions could be discontinued after 30 months was also terminated early because the children who were randomized to the no-transfusion group reverted to high-risk category (Mazumdar et al., 2007). Although no randomized clinical trials were conducted in the adult population to examine the benefits of continuing transfusion therapy for adults who had strokes as children, the evidence suggests that the brain is at risk for continued ischemic injury once a first stroke has occurred (Scothorn et al., 2002). Therefore, the benefits of routine screening and treatment in the pediatric population can only be successfully integrated into the adult system when efforts are taken to create systems and services to facilitate effective transition and transfer of patients to the adult healthcare system. Individuals undergoing regular treatment with simple chronic or exchange transfusion require uninterrupted medical care in order to decrease the morbidity and mortality associated with sickle cell disease.

Multiple blood transfusions are not without risk and can cause life-threatening complications to vital organs like the heart, liver and endocrine glands. One unit of red blood cells contains 200 to 250 mg of elemental iron (Gujja et al., 2010). Ballas (2001) reported an increased mortality rate of 64 percent versus 5 percent among patients with SCD who had iron overload. The prevalence rate of iron overload among the patients was as high as 33 percent (Ballas, 2001). The body does not have a mechanism to excrete excess iron, so repeated blood transfusions can result in iron overload. Individuals who have SCD and are either on chronic transfusion therapy or who receive frequent intermittent transfusion require iron chelation therapy (Kwiatkowski & Cohen, 2004; Wanko & Telen, 2005).
Excess iron acquired from red blood cell transfusion is deposited primarily in the cells of the liver, heart, pancreas and endocrine system. Fibrosis, cirrhosis and hepatocellular carcinoma are long-term outcomes of excess hepatic iron (Jensen et al., 2003; Kew, 2008). A study done by Darbari and colleagues (2006) found that 43.8 percent of those with iron overload had cirrhosis of the liver. Iron deposits in the heart is associated with myocardial stiffness, diastolic dysfunction and reduced left ventricular function. Individuals with myocardial iron toxicity can experience arrhythmias, and/or refractory heart failure as early as the second decade of life (Wood et al., 2005).

The benefits of chronic transfusion therapy to prevent strokes in patients with elevated jet velocity on transcranial ultrasound is dependent on iron chelation adherence. Therefore, in order to reduce the negative health outcomes associated with untreated iron overload, it is of critical importance that medical care is uninterrupted. This type of therapy requires close surveillance of not only iron chelation medication adherence but also regular assessment for organ damage so that treatment can be implemented in a timely manner.

Individuals who receive treatment for iron toxicity with chelation therapy reduce the burden of iron to vital organs. They also have improved survival (Shander & Sazama, 2010). If effective transition pathways are not in place to facilitate the uninterrupted transfer of care from the pediatric to the adult healthcare system, then individuals who receive transfusion therapy in childhood will either age out of care, get irregular care when symptoms are unbearable or experience complications that will result in increased morbidity and mortality.
2.2.6. **Bone and Joint**

Bone pain is the most common clinical manifestation of sickle cell disease. Vaso-occlusive crisis, which is occlusion and inflammation of the small blood vessels, is the most frequent complication of sickle cell disease that requires hospitalization (Platt et al., 1991). Although occlusion of the vessels with sickled cells can occur in any organ, it occurs most often in the bone marrow. Other complications involving the bones and joints are dactylitis (seen only in children under the age of 7 years), bony infarctions, osteomyelitis and septic arthritis, and avascular necrosis (AVN) (Almeida & Roberts, 2005). Avascular necrosis of the femoral head is the most common and disabling of the bony complications. Bone and joint complications can occur at any location in the body and are the most common cause of chronic pain for individuals with SCD (Aguilar et al., 2005).

Sickled cells cause occlusion of the small vessels, which leads to stasis and hypoxia. The ultimate result is infarction in the surrounding bone caused by ischemia. The bones most commonly affected are the long bones of the extremities, the spine, ribs and sternum (Khedr et al., 2012; Smith, 1996). In a study done by Khedr and colleagues (2012) using whole body magnetic resonance imaging (MRI) to assess musculoskeletal abnormalities in a population of 27 individuals with SCD, they found that all the patients had multiple bone infarcts. Eighty-five percent of them had vertebral infarction and 33 percent had AVN of the femoral head. Their findings were consistent with findings by Adekile and colleagues (2001) who reported that approximately half of all patients with SCD develop osteonecrosis by the age of 25 years (Adekile et al., 2001). Femoral osteonecrosis often leads to total collapse of the bones in the hip; this results in
debilitating pain that oftentimes requires hip replacement. Analysis of data from CSSCD reported by Milner and colleagues, states that 80 percent of those who underwent total hip replacement were younger than 35 years (Milner et al., 1991). Incidence of complications such as these highlight the importance of creating effective pathways for transition from pediatric to adult healthcare for those with SCD.

2.2.7. **Gastrointestinal and Hepatic**

Sickle cell disease affects many organs in the abdomen; the spleen, liver and gallbladder are the major organs that suffer the greatest insult from complications of the disease. The phenotype of the disease dictates the type of complication that occurs with the spleen. The splenic complication that is most common among individuals with Hgb SS is occlusion of the small blood vessels with sickled cells. The resultant infarction of the organ causes the spleen to atrophy. Individuals who experience this complication have increased susceptibility to infection from encapsulated bacteria such as *Streptococcus pneumoniae* and *Haemophilus influenzae* due to the non-functional spleen (Booth et al., 2010).

Splenic sequestration is another complication that occurs in infancy in all phenotypes of sickle cell. However, it can occur at any age among those individuals with Hgb SC and SC-thalassemia. Splenic sequestration is the pooling of blood in the spleen. The rapid pooling of blood in the spleen can result in hypovolemic shock and death within hours if medical intervention with blood transfusion is not undertaken in a timely manner (Ebert et al., 2010).
The liver is affected by both acute and chronic complications. Sequestration of blood in the liver is caused by obstruction of vessels by sickle cells. This results in liver enlargement and decreased hemoglobin. Once the obstruction is resolved, the trapped RBCs return to the circulation. This causes a rapid rise in the hemoglobin, which can result in complications such as heart failure and bleeding in the brain (Lee & Chu, 1996). Over time, multiple small infarctions that occur as a result of microvascular obstructions eventually lead to fibrosis of the liver. As previously stated, iron accumulation in the liver from transfused blood is another major complication that causes significant morbidity and mortality. Cirrhosis of the liver was found in 11.3 percent of individuals who had iron overload (Darbari et al., 2006; Ebert et al., 2010).

It is estimated that 26 to 58 percent of individuals with SCD will develop gallstones or cholelithiasis. Gallstones oftentimes cause obstruction of the common bile duct, which can initiate a cascade of other complications in the liver. This includes cholecystitis and biliary tract obstruction. Cholecystectomy is the most frequently performed surgical intervention for patients with SCD. It accounts for almost 40 percent of surgical procedures (Ebert et al., 2010).

Individuals undergoing surgical procedures require meticulous perioperative preparation in order to decrease the complications and mortality associated with surgery. It is estimated that approximately 7 percent of all deaths among individuals with SCD are related to surgery (Vichinsky & Lubin, 1980). The current standard of care for individuals with SCD who are undergoing major surgical procedures is to perform exchange blood transfusion in order to minimize sickling and to decrease Hgb S to 30 percent or less (Ingle & Ubale, 2011). Individuals who receive episodic care for pain
crisis are at risk for presenting sicker and for experiencing complications that would otherwise be identified and addressed earlier if they were in care.

2.3. Current Therapies

Although sickle cell disease is one of the most common inherited disease globally, there were minimal advances in the development of therapeutics until 2017. Treatment for SCD was limited to the management of symptoms, such as narcotic and anti-inflammatory medications for pain crises, and blood transfusion for symptomatic anemia, stroke, and acute chest syndrome. It is hypothesized that funding for sickle cell disease research and development lagged behind that of other genetic diseases like cystic fibrosis partly as a result of racial bias (Strouse et al., 2013). However, in 2017, the American Society of Hematology (ASH) partnered with federal agencies such as the National Institutes of Health to set priorities for sickle cell disease research, improve access to treatment, training and education of health care professionals (Gardner, 2018). The passage of the Sickle Cell Disease and Other Heritable Blood Disorders Research, Surveillance, Prevention and Treatment Act of 2018 provides grants for research, treatment, prevention and monitoring (Gardner, 2018). This legislation and commitment by the American Society of Hematology have led to significant advancement in sickle cell disease.

The major goal of blood transfusion is to improve the ratio of sickle cells to normal red blood cells. This results in the reduction of Hgb S concentrations and improved capacity of the red blood cells to deliver oxygen to organs. However, complications of transfusion such as increased viscosity of the blood can impede oxygen
delivery and worsen vascular occlusion (Styles & Vichinsky, 1994). Iron overload and red cell alloimmunization (formation of antibodies against own antigens on red cells) are also frequent complications of repeated transfusions (Chou, 2013).

Hydroxyurea (HU), an oral chemotherapeutic agent, has been the mainstay drug that is used to reduce and prevent many complications of SCD and improve quality of life. It works by increasing the concentration of fetal hemoglobin (Hemoglobin F - Hgb F) in the blood. Increased levels of Hgb F alter the process and prognosis of SCD. Increased levels of Hgb F have also been identified as a major positive prognostic factor for several of the complications of SCD. The presence of increased Hgb F causes a reduction in the number of white blood cells (WBCs), platelets and reticulocyte count. As a result, there is a reduction in the stickiness of red blood cells to the vascular endothelium and this reduces vascular occlusion (Voskaridou et al., 2010; Charache et al., 1995). The use of HU also decreases the frequency of painful crises, the frequency of hospitalizations, the incidence of acute chest syndrome as well as the death rate among those who use the drug (Hankins et al., 2005; Steinberg et al., 2003). Hydroxyurea also reduces vascular injury, which causes progressive organ damage as a result of sickling-induced hypoxia and infarction (Platt et al., 1994; Voskaridou et al. 2010). The use of hydroxyurea decreases the need for red blood cell transfusions. Overall, individuals taking HU usually have fewer painful crises and are at decreased risk of developing early and long-term complications of the disease (Charache et al., 1995).

Red blood cell transfusion has been an established treatment modality for both acute and chronic complications of sickle cell disease. Simple transfusion is administered in situations when a patient has symptoms (such as shortness of breath, light
headedness on standing) as a consequence of anemia. Chronic transfusion is done to prevent further damage to an affected organ (Telen, 2001). Multiple complications are associated with transfusion therapy; they include iron overload and hemolytic reactions (Darbari et al., 2006; Vichinsky et al., 1990).

Hematopoietic stem cell transplantation from an HLA-matched sibling donor offers the best chance for a cure. Transplantation eliminates the sickle red cells, thereby reversing the disease. However, this treatment option is underutilized for many reasons (Buchanan et al., 2010). Among them are the unavailability of a HLA-identical sibling donor and the belief that hematopoietic stem cell transplant is risky and carries a higher mortality rate than if transplant did not occur. However, studies have shown that the overall survival and event-free survival exceeds 90 percent. In one study that was done in New York, all 18 children survived with no evidence of sickle cell disease after hematopoietic cell transplant (Lucarelli et al., 2014; Bhatia et al., 2014; King et al., 2015).

Since 2017, major scientific progress has been made in the development of treatments for sickle cell disease. Two new drugs, Voxelotor and Crizanlizumab were approved by the Food and Drug Administration (FDA) in November 2019 (Aschenbrenner, 2020). Several clinical trials are underway investigating the use of gene therapy. Gene therapy involves the transfer of genetic material into a patient to treat his/her disease. The goal of gene therapy for genetic diseases like sickle cell is that the transferred gene when infused into the patient will be expressed at levels high enough that complications of the disease will be halted (Anguela & High, 2019). These modified stem cells will either correct the defective red cell mutation or induce Hgb F. Either of
these approaches, if successful, should result in longer life span of sickled red blood cells or eliminate the production of sickle cells and thereby eliminate the deleterious complications that are experienced (Orkin & Bauer, 2019).

2.4. **Comprehensive Sickle Cell Centers**

Sickle cell disease was identified by James Herrick in 1910; however, the translation of this knowledge into improved patient care lagged behind for several decades. This is partly a result of the intersection of race and stigma. In 1934, it was reported that black infant mortality rate in Memphis was 11 percent. However, it was not until the 1950s and 60s, when sickle cell disease was reframed as a health disparity as evidenced by high infant mortality for children with sickle cell disease under the age of two years (Siddiqi et al., 2013). Factors that converged to propel sickle cell disease from clinical obscurity into the socio-political landscape were not limited to the attention to racial inequality in health care, but it was also symbolized in movies with prominent African American celebrities like Sidney Poitier (*A Warm December*, 1974, in which he played a physician who loves a woman with sickle cell disease), the activism of the Civil Rights Movement of the 1960s and of civil organizations like the Black Panther. It is within these contexts that sickle cell disease was brought to national consciousness. As a result, in 1971, President Nixon included sickle cell disease as a national priority in his “Health Message” to the nation. In this message, President Nixon stated that, “It is a sad and shameful fact that the cause of this disease has been neglected throughout our history. We cannot rewrite this record of neglect, but we can reverse it.” As a result, the
92nd Congress enacted the National Sickle Cell Anemia Control Act of 1972 and President Nixon signed it into law (Wailoo, 2017; Siddiqi et al., 2013).

The National Sickle Cell Anemia Control Act paved the way for government funding for diagnosis, prevention through voluntary sickle cell screening and genetic counseling, research and treatment of sickle cell disease (Smith et al., 2006; Olney, 1999). As a result of the legislation, Comprehensive Sickle Cell Centers were established in 1972 to provide support for multi-disciplinary programs of basic, clinical and behavioral research; for core resources such as laboratory and data analysis; and for quality service activities including diagnosis, counseling and education (NHLBI, 1995). However, despite the allocation of Federal grants to develop comprehensive sickle cell centers, disparities in clinical care and research efforts still existed. It is within this historical backdrop that the Sickle Cell Treatment Act of 2004 was passed. This legislation expanded sickle cell disease specialized treatment centers that had been established by the prior legislation.

Since 1990, the State of New Jersey has included screening for SCD as part of the neonatal panel for genetic disorders. Approximately 80 – 90 infants are born with sickle cell disease in New Jersey annually (New Jersey Department of Health and Senior Services [NJDHSS], 2017). Immigration patterns to the United States is also impacting the incidence of sickle cell disease. New Jersey is among the states with the highest number of immigrants (Krogstad & Keegan, 2014). Immigration trends have the potential for adding more individuals with sickle cell disease to the population. In 2017, census data revealed that 5.7 percent of new immigrants were from Africa, 46.7 percent from
Latin America and 33.3 percent migrated from Asia (Batalova et al., 2018). These are the regions that have the highest number of individuals with both sickle cell disease and sickle cell trait.

In order to ensure adequate access to care, and to fulfill the requirements of sickle cell disease legislation, the New Jersey Department of Health and Senior Services (DHSS) developed a comprehensive model of referral and health care services whereby infants who screen positive for any abnormal hemoglobin are referred to one of seven comprehensive sickle cell disease care centers in the State for specialty care. In complying with the federal legislation, New Jersey DHSS provides funding through grants in one-year cycles to designated sickle cell centers in an effort to ensure access to specialty care for children with SCD. Although most children with SCD in the state receive SCD specialty care from these centers, there are other health care providers and hospital systems that provide care to pediatric patients with SCD. The latter do not receive targeted funding for the provision of comprehensive, family-centered care to this population. The number of children receiving health care services outside of the state SCD centers is unknown. Children with SCD who migrate to the United States are oftentimes referred to these SCD centers by their pediatricians. Sometimes they are connected to care by default, that is, they are admitted for SCD complaints to one of the hospitals within the catchment area. Once the pediatric hematologist admits the patient or consults for the management of SCD, when the child is discharged from the hospital, instructions are provided for follow-up in a sickle cell center, which is most often the one that is closest to where the patient/family resides.
The New Jersey State-funded comprehensive sickle cell centers are located at St. Joseph’s Hospital in Paterson, Rutgers Cancer Institute of New Jersey (R-CINJ) and its affiliate Bristol Myers-Squibb Children’s Hospital at Robert Wood Johnson University Hospital in New Brunswick, University Hospital in Newark, Beth Israel Medical Center in Newark, Hackensack University Medical Center in Hackensack, Cooper University Hospital in Camden, and Children’s Hospital of Pennsylvania (CHOP) at Voorhees campus. The pediatric sickle cell center at R-CINJ and the center located in Paterson provide care to the largest number of patients in New Jersey. R-CINJ also has a satellite clinic at Monmouth Medical Center in Long Branch, New Jersey.

The Sickle Cell Treatment Act of 2004, the Patient Protection and Affordable Care Act of 2010, as well as the objectives in Healthy People 2020, offer a remarkable opportunity to improve the health outcomes of individuals with SCD. The goals of these laws, as well as the objectives of the national health care agenda, stipulate that less-than-optimal availability and access to healthcare services, which are major determinants of health, is detrimental to living longer lives free of preventable diseases and disabilities.

Pediatric SCD care in New Jersey is organized in a family-centered, comprehensive, multidisciplinary manner. This care model creates a unique opportunity to include transition planning as an added element to the current comprehensive care model. Specifically, almost all the pediatric centers are located within larger tertiary health care systems. These facilities have the medical infrastructure to create transition from the pediatric to adult care because adult and pediatric hematologists (sickle cell disease specialists) are oftentimes located within a short distance of each other,
sometimes within the same building. However, pathways from pediatric to adult health care services for young adults with SCD are not consistent or comprehensive. The past ten years have seen exciting scientific innovations and dramatic improvement in SCD care; however, there is a major gap in the efforts to ensure that comprehensive, family-centered care is available to young adults as they move from pediatric to adult health care system.

2.5. Transition

Advances in medical technology, improvement in the diagnosis and medical management, and an improved health delivery system have resulted in increased life expectancy among adolescents and young adults with chronic conditions (Blum, 1995; Perrin et al., 2007; van der Lee et al., 2007). It is estimated that 21 percent of the US adolescent population has special health care needs and each year approximately 750,000 of these adolescents enter adulthood (Scal & Ireland, 2005). Increased life expectancy has presented new challenges for these young adults. It is expected that by the age of 18 to 21 years individuals with pediatric onset chronic diseases should be receiving age-appropriate care in the adult healthcare system (Reiss et al., 2005). Despite this expectation, pediatricians continue to provide care to young people with chronic diseases well into the adult years. Transfer usually takes place haphazardly, with the most common options being lack of continuity of care and reliance on emergency department services, abrupt transfer to the adult system, or falling through the ‘gap’ with significant adverse health consequences (Boyle et al., 2001; Fleming et al., 2002). Efforts to improve the provision of transitional services have resulted in policy statements on transition by
professional organizations, policymakers, healthcare practitioners, and healthcare delivery systems. However, the system still continues to struggle to provide timely, comprehensive transition services.

In 2002, several professional organizations convened and put forth a consensus policy statement on transition. The American Academy of Pediatrics, the American Academy of Family Physicians, and the American College of Physicians-American Society of Internal Medicine panel have identified six critical steps to ensure that effective transition occurs:

1. All youths must have an identified healthcare provider that is addressing transition issues
2. Transition training must be incorporated into physician competencies
3. Patients must have portable medical records
4. Each patient must have an individualized transition plan
5. Youths with special health care needs must have access to primary and preventive services
6. They must have access to affordable and continuous health coverage (AAP, 2002a)

Provisions in the Patient Protection and Affordable Care Act (ACA) of 2010 addressed elements that would facilitate improved access to health insurance coverage, preventive services and coordination of services (Shaw et al., 2014). Despite these guidelines and provisions, there are struggles within New Jersey’s current health care system to define and develop successful, comprehensive transition services for adolescents and young adults with chronic illnesses like SCD.
The American Society for Adolescent Medicine describes transition as an active medical process, “the purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from a child-centered to adult-centered healthcare systems.” (Blum et al., 1993). In 1993, Blum and colleagues argued that the goal of transition planning is to provide comprehensive, continuous health care that is developmentally appropriate and that meets the psychosocial and family needs of adolescents and young adults with pediatric onset diseases. A more comprehensive definition of transition is “a multifaceted, active process that attends to the medical, psychosocial, and educational-vocational needs of adolescents as they move from child oriented to adult oriented lifestyles and systems” (White, 1997). Although the chronological age at which transition should begin varies in the literature (from 11 years to 20 years), there is consistency in that the timing should be guided by such factors as developmental readiness and health status (Viner, 2001; McDonagh et al., 2000a). It is clear in the literature that transition is a process that should be initiated years before the official transfer to the adult system occurs (Blum, 1995; Viner, 2001; Reiss & Gibson, 2002; Fox et al., 2002; Sawyer et al., 1997). The transition process should include education about the disease, acquisition of self-management skills like scheduling appointments and taking responsibility for medication administration, having the ability to communicate with healthcare providers and having clear timelines for acquisition of those skills (Blum, 1995; Rosen, 1994).

Transition in terms of adolescents and young adults with SCD refers to the uninterrupted movement from pediatric to adult health care services. The transition process occurs over time and ultimately culminates in the medical transfer of care from
the pediatric to the adult health care system. Since most pediatric patients with SCD receive their treatment within designated comprehensive sickle cell centers, it would be advantageous to develop and implement standardized processes to address the problem of ineffective transition within these centers. This problem is not unique to patients with SCD, many studies focusing on young adults with pediatric onset chronic diseases have revealed that the vast majority do not receive adequate preparation to allow them to competently navigate the adult health care system. In 2004, Yankaskas and colleagues found that 20 percent of adults with cystic fibrosis were receiving their medical care in pediatric settings (Yankaskas et al., 2004). In the 2001 National Survey of Children with Special Health Care Needs, only 15 percent of those who were interviewed (N = 5400) reported that they received appropriate transition preparation to make the move to the adult system (McPherson et al., 2004). Maintaining optimal health is crucial for individuals with pediatric onset chronic conditions like SCD; therefore, it is critical that there is seamless transition of care from pediatrics to adult health care services.

The life expectancy for adults with SCD is 42 years for men and 48 years for women, and many live into the fifth and sixth decades (Platt et al., 1994). These individuals require life-long surveillance in order to decrease morbidity and mortality and improve their quality of life. A single-center cohort study conducted by Quinn and colleagues (Quinn et al., 2010) found that majority of deaths that occurred in individuals over the age of 18 years occurred after transfer to an adult provider. Other studies have also found that patients experience worse outcomes such as non-adherence to immunosuppression medications after liver transplant and increased hospitalizations for
patients with diabetes after they transition to adult care (Annunziato et al., 2007; Nakhla et al., 2009).

According to a statistical brief from the Healthcare Cost and Utilization Project sponsored by the Agency for Healthcare Research and Quality, and the results of a retrospective cohort of SCD-related emergency department visits and readmissions from eight geographically dispersed states, sickle cell disease had the highest 30-day readmission rates for individuals aged 18 to 30 years old. That is more than that of any other medical condition (Brousseau et al., 2010b; Elixhauser & Steiner, 2013). These findings highlight the crucial need for patients to have effective transition from pediatric to adult healthcare system. Long-term patient surveillance is critical for the detection, treatment and prevention of acute and chronic complications of sickle cell disease. This can only be accomplished through the development of comprehensive transitional health care models that are planned and coordinated. Unsuccessful transitioning will force young adults to be discharged from pediatric care without the appropriate skills that are needed to navigate the adult health care system. Skill sets that are needed include the scheduling of appointments and keeping them, knowing their medical history, being able to identify signs and symptoms of illness that require immediate medical attention and being able to advocate for one’s self during medical encounters. Although they may ultimately end up in the adult system, the lack of knowledge and skills needed to manage their disease puts them at risk for negative outcomes. The lack of, or deficiency in, the necessary skills to negotiate the adult health care system may result in them returning to the pediatric provider, using the emergency department for health care services, or even falling out of care (Hauser & Dorn, 1999; Wojciechowski et al., 2002).
The organization of care in both systems may be a major factor for the delay in transition from the pediatric to adult system. Patients with pediatric onset chronic conditions usually view their pediatric specialist as their primary provider because that is where they receive treatment and preventive services in a comprehensive, coordinated manner (Lanzkron et al., 2018). A survey done by Okumura and colleagues (2008) found that a majority of internists reported that they were uncomfortable in providing care to young adults (ages 17 to 25 years) who had pediatric onset chronic diseases.

Approximately sixty percent of pediatric patients with SCD have public insurance (Brousseau et al., 2010a). The National Health Interview Survey in 2008 demonstrated that twenty-nine percent of young adults aged 18 to 24 years were uninsured. This is the highest uninsured group compared to any other age group (Hunt & Sharma, 2010). Underinsurance and lack of insurance presents major challenges for young adults with chronic conditions like SCD. The Patient Protection and Affordable Care Act (ACA) of 2010 provides benefits such as allowing young adults to remain on their parents’ insurance until the age of 26 years old. The cost-sharing provision of the ACA also provides the opportunity for insurance coverage to the uninsured thus eliminating the decreased access to health care services. Despite these provisions, the process of providing the necessary documentation to prove eligibility annually can be burdensome, especially for someone who has had strokes, that many of these young adults remain in the pediatric system where they have access to social services and where their care is delivered in a comprehensive, coordinated way. Other system barriers that young adults confront include inadequate financial reimbursement to providers for care coordination and lack of institutional support for transition services (Geenen et al., 2003; McDonagh et
al., 2000b; Scal & Ireland, 2005; Scal, 2002). System barriers such as inconvenient office
hours and lack of qualified or knowledgeable professionals in the adult system have also
been identified as reasons why young adults are still receiving care in the pediatric setting
(Scal et al., 2008).

Barriers have also been reported from several perspectives: patients, parents and
caretakers, pediatric health care providers and adult health care providers. Patient barriers
include the reluctance to leave the safety of the pediatric providers. Hauser and Dorn
(1999) conducted separate focus groups among adolescents, parents and providers and
showed that adolescents and their parents fear leaving the care of the familiar physician
and going to an adult physician who was not familiar with SCD. Limited or inadequate
knowledge about their health condition, unpreparedness about the structure and care
delivery in the adult system, lack of trust of providers in the adult system, and fear of
leaving the familiar pediatric providers and health care setting are some of the reasons
why young adults still receive care in pediatrics (McPherson et al., 2009; Telfair et al.,
1994).

Parents and caretaker barriers include concerns that young adults will not be able to
manage their care. Parents also report that they perceive that adult providers do not want
them to be involved in the care of their children (Telfair et al., 1994).

Pediatric healthcare provider barriers include difficulty relinquishing care because
they have provided care all or most of their patients’ lives, lack of the knowledge and
skills of adult providers, shortage of available adult health care providers with the
knowledge and skills to provide care to these young adults, lack of reimbursement for
transition services and inadequate time to deliver transition services (Nehring et al., 2015; McManus et al., 2008).

Adult healthcare provider barriers include lack of interest in taking care of young adults, especially if they have multiple complications such as strokes. The adult healthcare system is more rapid-paced and patients who have complications require more time for an office visit. Adult providers are not accustomed to dealing with parents and oftentimes have concerns about confidentiality and HIPPA violation (Peter et al., 2009). Research shows that adult healthcare providers have identified a number of barriers to transitioning young adults with pediatric onset chronic conditions like sickle cell disease. These include inadequate time to provide the required service, inadequate resources, inadequate knowledge about how to provide care to individuals who may have experienced a neurological insult (stroke) as a child, and lack of understanding of the patient’s situation (Collins et al., 2012; Okumura et al., 2008).

Health care system barriers include the lack of formalized transition programs (Fernandes et al., 2012), problems with getting adult specialty care, poor communication between adult and pediatric providers (Collins et al., 2012), and differences in the organizational structure of pediatric and adult systems of care (Kingsnorth et al., 2011; Reiss et al., 2005).

2.6. Models of Transition

Several models of transition have been proposed for children with chronic illnesses like sickle cell disease. The disease specific model uses a transition approach where individuals move from a pediatric specialist to a team with pediatric and adult
specialists, and then eventually to the adult system. The *generic model* of transitional care is provided in a developmental manner; the patient moves from pediatric to adolescent, and then to adult health services. The *primary care model* uses the family practitioner or general practitioner to coordinate care, and referrals are made to specialists on an as needed basis. The *single-site model* uses the same clinical site and is similar to the generic model: the patient moves from pediatric to adolescent and then to adult care within the same health system (Ginsberg et al., 2006; Henderson et al., 2010). Programs that use these models of care are not developed within a conceptual framework. This makes it difficult to formulate and implement a best practice template for healthcare transition across disease states in adolescents and young adults with chronic diseases.

The standard of care for individuals with sickle cell disease is prevention of complications by close surveillance by the health care team, prompt medical attention when disease challenges like painful crises, fever, sudden onset of neurological symptoms arise, and regular health maintenance that includes routine screenings and immunizations. A review of the transition literature suggests that transition is oftentimes haphazard with no clear blueprint for best practices. It is through these lens that the issue of transitioning young adults with sickle cell disease from pediatric to adult health care setting is examined.

### 2.7. Theoretical Foundation

Sickle cell disease is a complex disorder that affects multiple organs in the body. Young adults with the disease are at risk for experiencing gaps in medical care as they age out of pediatrics (Betz, 2004; Viner, 1999). Therefore, a multifaceted approach is
crucial for successful transition and should be guided by a conceptual framework. The major benefit of using a conceptual framework is articulated by the American Academy of Pediatrics (AAP) in its policy statement that all children and youth with special health care needs should have a medical home (AAP, 2002a).

In this study, the researcher utilized concepts of the Patient-Centered Medical Home Model and elements from the National Quality Forum Framework for Measuring Care Coordination as guide to frame and develop the questions that were used to answer the main research question of why young adults with sickle cell disease continue to receive healthcare services in the pediatric setting. The Patient-Centered Medical Home (PCMH) model, designed around the needs of the patient, is defined as the “provision of comprehensive primary care services that facilitates communication and shared decision-making between the patient, his/her primary care providers, other providers, and the patient’s family” (American Hospital Association, 2010). The PCMH is designed around the needs of patients with chronic diseases. The primary goal of this model is to improve access to quality comprehensive routine care through coordination of care and effective communication among all members of the health care team, and availability and utilization of appropriate preventive health care services (Jackson et al., 2013). It also advocates for the use of health information technology to collect data, and for effective communication between patients and health care providers (Weedon et al., 2012). The management of sickle cell disease is complex for most individuals, so achieving these goals will undoubtedly result in decreased morbidity and mortality, as well as improved quality of life. Prevention and/or delay in onset of complications like strokes and pulmonary hypertension, less use of emergency department, and fewer hospital
admissions requires a comprehensive approach, and the PCMH model fulfils this need.

Ferrante and colleagues (2010) outlined six interrelated elements that are important for successful transition to occur:

1. accessible care: physically accessible, convenient operating hours, and the ability to schedule appointments.
2. continuity of care: having continuous care without any gap in services
3. coordination of care: coordination of care with the multidisciplinary team, as well as good communication between and among providers, patient and/or family
4. comprehensiveness: collaboration of the multidisciplinary team, patient, and/or family
5. quality and safety: use of evidence-based guidelines and clinical decision-support tools
6. use of information technology: accessible electronic medical records

The complexity of disease management and the need for multi-disciplinary services made using elements of the PCMH framework essential in framing the questions that provided the answers to the larger research question.

The National Quality Forum Framework for Measuring Care Coordination describes the basic elements needed for optimal transition of care in terms of structural quality, the common processes that should occur in any setting of care and that can be applied to all patients experiencing transition. The NQF describes health care coordination as a process “that ensures that patients’ needs and preferences for health services are met.” It also advocates that there should be effective communication between the patient and family, health care provider, proper health maintenance (surveillance and
prevention) and facilitation of information sharing across health care disciplines and sites (NQF, 2006). Its focus is on effective care coordination across all settings in order to prevent gaps in care, especially for individuals with chronic conditions like sickle cell disease. Effective communication and coordination of care are two of the major pillars for successful transition (National Quality Forum, c2021). Consequently, elements of both frameworks were used to guide the design of the interview guide for patients and the survey for providers in both the pediatric and adult healthcare systems. This comprehensive approach provided an opportunity to understand the issues that are critical to timely, successful transition for young adults with sickle cell disease.
CHAPTER THREE

RESEARCH METHODS

3.1. Research Questions

1. What are patients’ perceived system, individual and provider factors that facilitate and/or impede transition from pediatric to adult health care services for young adults with sickle cell disease?

2. Are young adults with sickle cell disease being prepared to access and navigate the adult health care system?

3. What are the health care providers’ perceptions of patient, system and providers’ factors that impact transition from pediatric to adult services?

4. How and when should transition from pediatrics to adult health care services begin?

3.2. Overview of the Study Design

Successful health care transition remains a major challenge for adolescents and young adults with sickle cell disease. Studies have shown that when patients with sickle cell disease cross the threshold from childhood to adulthood, they either remain in the pediatric system longer than they should or leave pediatric care unprepared to navigate and utilize sickle cell disease preventive healthcare services (Telfair et al., 1994; Hauser & Dorn, 1999; Wojciechowski et al., 2002). This chapter summarizes the methods that were used to answer the research questions.

The issue of transition from pediatric to adult health care services for individuals with chronic illnesses like sickle cell disease is very complex. A mixed-method research design was used to explore why young adults with sickle cell disease are still receiving
health care services in pediatrics. This research design allowed for the simultaneous collection of both qualitative and quantitative data and their integration to identify common threads necessary to understand the problem. This approach enabled the researcher to collect richer and stronger evidence that could not have been accomplished by using a single method.

The qualitative data gathered through individual interviews and memos provided detailed information about the problem by emphasizing the voices of patient participants. The quantitative portion gathered descriptive information from health care providers regarding the current state of medical care and their views of barriers and/or facilitators for seamless transition for this population of patients. The use of this mixed method approach allowed the researcher to get in-depth information about the current state of transition, the process of transition, those factors that facilitate and/or impede transition, and the elements necessary to ensure that transition occur in a planned and coordinated manner. This approach also allowed for intense exploration of the problem using a small number of subjects in their natural setting (Creswell, 2002; Yin, 2009). Figure 1 outlines the study design.

![Figure 1: Outline of Study](image-url)
Table 1 provides details of the study design, including the target population, the purpose for collecting data from each population, the time period over which data was collected, the sample size, data collection methods and the link to each research question.

**Table 1: Overview of Research**

<table>
<thead>
<tr>
<th>Target Population</th>
<th>Purpose</th>
<th>Time Period</th>
<th>Sample Size</th>
<th>Data Collection Method</th>
<th>Link to Research Question</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patients (P)</td>
<td>To better understand the state of transition and the factors that impact transition from pediatric to adult system</td>
<td>April 2016 to November 2017</td>
<td>N = 16</td>
<td>Individual interviews</td>
<td>1, 2 &amp; 3</td>
</tr>
<tr>
<td>Health care Providers (HCP)</td>
<td>To understand the challenges clinicians encounter, to identify system, patient and provider level factors that impact transition</td>
<td>March 2017 to October 2017</td>
<td>N = 38</td>
<td>Electronic web-based survey</td>
<td>3</td>
</tr>
<tr>
<td>Patients &amp; HCP</td>
<td>To determine the timeline for transition, to identify strategies that could be helpful and to elicit recommendations for what would work</td>
<td>April 2016 to November 2017 &amp; March 2017 to October 2017</td>
<td>N = 54</td>
<td>Individual interviews (P) &amp; Open-ended questions on electronic web-based survey</td>
<td>4</td>
</tr>
</tbody>
</table>
3.3. The Research Setting

The patient and health care provider samples were recruited from the pediatric hematology clinic at the Rutgers Cancer Institute of New Jersey, the adult hematology clinic Rutgers University Medical Group, the physician arm of Robert Wood Johnson Medical School and from Robert Wood Johnson University Hospital in New Brunswick, New Jersey. The pediatric clinic is located in the Rutgers Cancer Institute of New Jersey, and the adult clinic is located in the Clinical Academic Building on the same campus. There are two separate emergency rooms, one that serves patients from birth to 21 years of age, and the other that serves patients over the age of 21 years. Both emergency rooms, pediatric and adult, are located at Robert Wood Johnson Barnabas Health University Hospital (RWJBH) on the same campus as the pediatric and adult hematology clinics in New Brunswick, New Jersey.

3.4. Component 1: Interviews of Patients with Sickle Cell Disease

The target population for the qualitative component was adolescents and young adults with sickle cell disease who were receiving sickle cell disease specialty care in the pediatric hematology clinic, the adult hematology clinic and those individuals who sought health care services from the emergency room for sickle cell disease related complaints, but who were not necessarily in steady care. Grounded theory guided theoretical sampling (N=24). This approach allows simultaneous collection and analysis of data, so that the researcher knows when there is no new information emerging. Purposeful sampling was best suited to recruit adolescents and young adults with sickle cell disease aged 18 to 30 for this study because the participants have the depth of knowledge about
their lived experiences as they age out of pediatric care (Creswell & Plano Clark, 2011). Eliciting information from this homogeneous sample provided much richness to the data.

The age range for the participants was chosen because it is more likely that those at the lower age limit would be actively preparing to transition to the adult system and those at the upper age limit would recall their experience of moving from the pediatric system to the adult system. Apart from the age ranges, only patients with Hgb SS, Hgb SC, and Sβ Thalassemia variants (Sβ thalassemia plus and Sβ thalassemia zero), those who spoke and understood English and had no intellectual impairment or mental disorders were eligible to participate in the study, as depicted in the figure below.

### Table 2: Patient Eligibility Criteria

<table>
<thead>
<tr>
<th>Sickle cell variant</th>
<th>Hgb SS</th>
<th>Hgb SC</th>
<th>Sβ Thalassemia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient characteristics</td>
<td>English speaking</td>
<td>English speaking</td>
<td>English speaking</td>
</tr>
<tr>
<td>Age 18-30 years</td>
<td>Age 18-30 years</td>
<td>Age 18-30 years</td>
<td></td>
</tr>
</tbody>
</table>

Table 2: Patient Eligibility Criteria

#### 3.4.1. Recruitment of Patient Participants

Approval for this study was obtained from the Research Utilization Group (RUG) Committee at Robert Wood Johnson University Hospital and from the Institutional Review Board (IRB) at Rutgers University. In compliance with IRB requirements, the researcher posted IRB-approved information flyers (Appendix 1) that explained the purpose of the study, the target population (individuals with sickle cell disease between the ages of 18 and 30 years old), how data would be collected (individual interview), the anticipated length of each interview (30-45 minutes), that information would be
confidential, that a $25.00 gift card would be given as an incentive at the completion of the interview and the contact information of the PI. These flyers were posted in the waiting room of the pediatric and adult clinics and in the nurses’ stations in the adult and pediatric emergency rooms. The researcher explained the purpose of the study to the registration clerks, physicians, nurse practitioners, physician assistants and staff nurses in both hematology clinics (adult and pediatric) and to the staff in the ER and solicited their assistance in recruiting participants. In qualitative research, gatekeepers are often used to assist the researcher in gaining access to the study population (Berg, 2004; Creswell, 2003). Since the health care providers and registration clerks had access to the age and diagnosis of the patients, they were asked to inform potential recruits about the study and hand them the information sheet that explained the details of the study. The researcher also made visits to the clinical areas outlined in the study at least twice monthly during the study period to replenish flyers and to remind the staff of the study and elicit their assistance with recruitment.

3.4.2. Informed Consent

The purpose of the study was explained to each participant prior to obtaining written consent (Appendix 2). The researcher explained that participation was entirely voluntary and that the subject could withdraw at any time. Participants were also informed of the risks and benefits of the study and that no identifying information would be used in the report. The only personal risk was the possibility for a breach of confidentiality, but they were informed that there were safeguards in place to prevent a breach. They were also informed that there would be no direct benefit to them for
participating in the study but that the information gained from the study could be beneficial in that it could provide information as to how to develop a successful transition program. After consent was obtained, each participant was assigned a pseudonym in the form of a numeric code to ensure confidentiality. All consents were obtained in-person by the researcher. Each participant was handed the advertisement flyer after consent was signed because the flyer had the contact information of the researcher.

3.4.3. Areas of recruitment, number recruited and number of participants (N = 16)

- Pediatric Clinic: Eight individuals were recruited, six signed consent, three subjects were interviewed, two did not return telephone calls and one subject did not have a working telephone.
- Adult Clinic: Four participants recruited, three signed consent and three were interviewed
- Pediatric Emergency Room: Two referrals, see other for further details
- Adult Emergency Room: No referrals were obtained
- Other: The researcher is employed as an Advanced Practice Nurse on the Pediatric Hematology/Oncology inpatient service in the Children’s hospital and so has access to patients who met the inclusion criteria. Eighteen patients were approached for participation in the study, fourteen signed consent and ten were interviewed (including the two from the pediatric Emergency Room).
3.4.4. Data Collection and Instrumentation

The researcher created a list of names and telephone numbers of the participants who had provided consent. This list was saved as a separate file that could not be linked to the interview data. It was also used to keep track of the dates on which the interviews were completed. This list was kept on the researcher’s personal computer, which requires access with a secure password. This was done to protect the privacy and confidentiality of the participants. A total of twenty-four (N=24) subjects signed consent forms. However, just sixteen participants (N=16) completed the interviews because 6 participants did not answer their telephones and 2 participants’ telephones were not working. The participants who completed the interviews were given a $25.00 Visa card to compensate them for their time.

For those individuals who agreed to participate when face-to-face contact was made and who signed consent forms at that time, the interview was immediately scheduled, and no further calls were made. For others who signed the consent form and were not able to schedule appointment at the time the form was signed, a follow-up telephone call was made to the participant within a one to two-week timeframe of obtaining consent to arrange for a convenient time for the interview. If there was no response after the first call was made, two additional attempts were made to contact the participant. The expediency of such contact within the timeframe was important in order not to lose someone who expressed interest in participating in the study. Despite attempts to schedule the interviews expeditiously, majority of interviews were rescheduled from the original dates that were decided.
3.4.4.1. **Interview Guide**

The semi-structured interview guide was developed based on a review of the literature and guided by concepts in the Patient-Centered Medical Home Model (PCMH) and National Quality Forum Framework (NQF) for Measuring Care Coordination (Jackson et al., 2013; Antonelli et al., 2009). The researcher therefore combined elements of both frameworks to develop the interview guide.

The interview guide was designed to:

1. explore patients’ perceptions regarding their accessibility to healthcare services
2. explore their understanding of continuity of care and coordination of care
3. examine patients’ experience with current care including having a usual place of care
4. explore patients’ perceptions of how transition from pediatric to adult services should occur.

To address these areas, the semi-structured interview guide was developed to explore the following domains:

a. self-rated health
b. understanding of sickle cell disease and medical complications of the disease
c. understanding of the treatment plan
d. experiences and satisfaction with current healthcare services
e. understanding of continuity of care
f. experience with coordination of care
g. preparation for self-care management
h. preparation for transition and transfer of care
i. comfort with communication between provider and patient
j. use of electronic communication including access to electronic medical record

The interview guide used open-ended questions that were grounded in the elements and the domains of the framework and were intended to elicit views consistent with the “lived experiences” of the participants. The guide also captured non-identifying demographic information such as age, employment status, education, gender, insurance status, place of care (pediatric/adult service) and sickle cell variant (Appendix 3).

3.4.4.2. Individual Interviews

In-depth, one-on-one interviews were conducted either in person or by telephone using the semi-structured interview guide. They were conducted in English and each interview lasted between twenty-five to forty-five minutes. Fourteen interviews were conducted by telephone and two were conducted face-to-face. Fourteen of the interviews were recorded using a digital voice recorder to ensure that the participants’ full accounts were captured. Two participants declined audio-recording, so responses were handwritten during the interview and later typed in a Microsoft Word document.

The interviews were informal, carried out in conversational style, and mainly used open-ended questions. The questions were designed to encourage participants to fully describe their perceptions regarding transition, preparation for transition, the transfer process, transition facilitators/barriers and transition needs. Patient participants’ knowledge of sickle cell disease and complications of the disease were also included in the interview. Although an interview guide was used to lead the discussions, the structure of the interview was flexible. This format enabled the researcher to pursue unexpected but relevant information that emerged during the interview. The researcher was able to
use probes to stimulate the respondents to elaborate or produce more information when necessary.

The use of open-ended questions enabled the researcher to get a more in-depth understanding of the transitional issues with which young adults with sickle cell disease contend. The interview guide also included items that explored access to sickle cell disease specialty care, continuity of care and use of information technology. Some questions also attempted to elicit information regarding participants’ perceptions of what might work to resolve the problem of transition. The structure of the interview (audio-recording, note-taking) and that no identifying information would be made available to anyone who was not directly involved with the study (except faculty advisors only) was also explained to each participant. All interviews were conducted in a private space.

3.4.4.3. **Data Management and Analysis**

The researcher utilized principles of Grounded Theory in conducting the analyses. Grounded theory is an inductive approach that is used to generate theory (how things work) about the experiences of a group of individuals who share a common problem (Pursley-Crotteau et al., 2001; Glaser, 1978). This methodology was introduced by Glaser and Strauss (1967) as a systematic approach to study social interactions geared at generating theory from the iterative simultaneous collection of data and the utilization of constant comparative analysis until theoretical saturation is achieved (Gephart, 2003).

After each interview, the sound files were uploaded to a computer and the quality of the recordings were checked and verified. The researcher listened to each recording before sending it to be transcribed. Audiotaped interviews were transcribed verbatim in
order to establish a permanent written record and to facilitate further analysis. All recordings were transcribed by a professional transcription service. The transcriptions were manually checked against the sound files and notes to verify accuracy. The transcribed data were transferred into QSR NVivo-12 software for data management and analysis. The transcripts were analyzed using the constant comparative method to generate themes and sub-themes relating to the research questions. Five concepts generated from a review of the literature was used to form the framework for the organization of the data.

1. Sickle cell disease (knowledge of disease, complications and need for surveillance)
2. Transition (facilitators, barriers, timing, readiness, preparation and experience with transition)
3. Access to health care services (pediatric care, adult care and experience in both systems)
4. Communication (provider to provider, patient to provider and vice versa)

This thematic, organizational framework helped to identify and categorize the factors that converged to impact timely transition from pediatrics to adult health care services for young adults with sickle cell disease.

3.4.4.4. Human Subjects

To ensure that confidentiality was always maintained, participant data did not include names, dates of birth or addresses. Numeric pseudonyms were assigned to all patient participants. The signed consent forms were the only thing that could link patient
participants to the study. However, consent documents were placed in a folder and kept in a locked cabinet in investigator’s office. The form used to collect data that included names, telephone numbers and dates when calls were made to patient participants to schedule interviews were kept in a separate place from the data itself. The demographic data had no identifying information that could be used to link the participants to the study. Extensive precautions were taken to protect the confidentiality of participants. Rutgers University School of Nursing and Robert Wood Johnson Medical School IRB and HIPAA regulations concerning confidentiality and information availability were strictly enforced. The PI’s office computer was protected from outside access by RWJUH firewall and personal laptop computer was password protected.

3.5. Component II: Survey of Health Care Providers

The second component of the research design was a survey of health care providers that included physicians, nurses and nurse practitioners from the pediatric hematology clinic; physicians and nurses from the adult hematology clinic; and physicians, physician assistants and registered nurses from the pediatric and adult emergency rooms. Physicians who rotated through the various clinical areas as residents or fellows were not surveyed because of their limited interactions with this patient population. Apart from professional titles (MD/DO, NP, PA, RN), inclusion criteria were, therefore, limited to regular full-time and part-time employees in the clinical areas stated above. These professionals were chosen because of their frequent encounters with patients with sickle cell disease.
Table 3: Health Care Providers (N=38)

<table>
<thead>
<tr>
<th>Health Care Providers</th>
<th>Pediatric Emergency Department</th>
<th>Pediatric Hematology</th>
<th>Adult Emergency Department</th>
<th>Adult Hematology</th>
</tr>
</thead>
<tbody>
<tr>
<td>MD/DO</td>
<td>x</td>
<td>x</td>
<td>x</td>
<td>x</td>
</tr>
<tr>
<td>RN</td>
<td>x</td>
<td>x</td>
<td>x</td>
<td>x</td>
</tr>
<tr>
<td>NP/PA</td>
<td>No NP/PA</td>
<td>x</td>
<td>x</td>
<td>No NP/PA</td>
</tr>
</tbody>
</table>

Table 3: Eligible health care providers

3.5.1. **Recruitment of Health Care Providers**

To recruit health care providers, the principal investigator met with the medical director of the pediatric hematology/oncology clinic, explained the details of the study and solicited his assistance in informing the health care providers in the pediatric hematology/oncology clinic and the adult and pediatric emergency department (ED) to participate in the study. A subsequent meeting was arranged with the medical director and the research director of the emergency department, the pediatric hematology/oncology director and the researcher. They all agreed to announce the study in their monthly staff meetings, and they also provided feedback as to the best way to inform their respective staffs. The researcher also met with the director of nursing research, the head nurses in the pediatric and adult EDs, the nurses in the adult and pediatric clinics to explain the details of the study and to solicit their assistance in announcing the study at their staff meetings, as well as to encourage the nurses to complete the survey because their input was important in understanding the problem.

Electronic mail was sent to the administrative assistant of each department (adult and pediatric hematology, pediatric and adult emergency departments) with the details of
the study. They were asked to disseminate the survey to the clinical staff (Appendix 4). This approach was chosen because the administrative assistant in each department had access to the departments’ list of providers’ emails. This approach was also convenient because the researcher did not necessarily need to know the identities of the health care providers. The email included an explanation of the study and a link to the survey using Survey Monkey (Appendix 5). Follow-up emails were sent after the initial email was sent to all providers asking them to complete the study if they had not already done so. The researcher also made face-to-face contact with some providers (nurses and physicians) in the adult and pediatric ED to ask for their participation in the study. The emergency department is a very busy clinical area and the researcher surmised that it was likely that the clinicians had little time to complete the survey. Consequently, the researcher decided to take the extra step of going to the department to ask providers in person to participate in the study. Limited demographic information was requested; no personal identifiers or medical information was asked of the health care providers.

3.5.2. **Informed Consent**

No written consents or waivers were obtained from health care providers; their decision to complete the survey was sufficient.

3.5.3. **Areas of Recruitment and Number of Participants (N=38)**

Individuals with sickle cell disease receive sickle cell disease care from hematologists (pediatric and adult). However, when they experience any symptoms of sickle cell, the emergency department is usually the first point of contact. As such, health care providers
(physicians, registered nurses, nurse practitioners, physician assistants) from the pediatric hematology clinic, the adult hematology clinic and the pediatric and adult emergency departments were recruited. The following is a chart depicting the participants and the departments in which they work.

**Table 4: Areas of Recruitment of Health Care Participants (N = 38)**

<table>
<thead>
<tr>
<th></th>
<th>Physicians</th>
<th>RN</th>
<th>NPs</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Emergency Department</td>
<td>7</td>
<td>15</td>
<td>0</td>
<td>22</td>
</tr>
<tr>
<td>Pediatric Hematology</td>
<td>5</td>
<td>6</td>
<td>4</td>
<td>15</td>
</tr>
<tr>
<td>Adult Hematology</td>
<td>0</td>
<td>1</td>
<td>N/A</td>
<td>1</td>
</tr>
</tbody>
</table>

- Emergency Department: **Seven** physicians, **two** RNs from the adult emergency department, **thirteen** RNs from the pediatric emergency department and no physician assistants
- Pediatric Hematology: **Five** physicians, **four** NPs and **six** RNs
- Adult Hematology: **One** RN and no physician

### 3.5.4. Data Collection and Instrumentation of Clinicians

The method used to identify the perceived factors that impact transition from pediatric to adult health care was a *survey*. An electronic web-based survey was conducted among health care providers (N = 38) who provide health care to patients with sickle cell disease. Advantages of using this method included low collection costs, the ability of the respondents to complete the survey at their convenience and the ability of
the researcher to gather numeric information on an instrument. Disadvantages of this data collection method are the potential misinterpretation of questions, so the answers provided do not necessarily capture the participants’ true feelings.

3.5.4.1. Survey development

Electronic web-based survey questions were guided by the elements of the National Quality Forum Framework for Measuring Care Coordination (Antonelli et al., 2009). The survey was designed to explore the structure, processes and outcomes of the existing system of care for young adults with sickle cell disease in the pediatric and adult settings. Thus, the design:

1. explored provider’s perception of accountability during all points of transition
2. examined whether there was a tool for plan of care
3. examined the care team processes (e.g., ability to access medical records and referrals to other providers)
4. explored providers’ perceptions of their experience with sending and receiving patients from pediatric to adult services, experience and satisfaction with the quality of interaction and collaboration between the pediatric and adult providers
5. explored providers’ perceptions about how transition from the pediatric to adult system should be designed and implemented.

The survey was also designed to explore the following domains because these elements are fundamental to successful transition: manpower for coordination of care, system factors that impact coordination of care, and use of information technology and its impact, positive and/or negative, on health care delivery. Additionally, the questions were organized around
the six core elements of the Health Care Transition model as outlined by the National Health Care Transition Center: transition policy, transition registry, transition preparation, transition planning, transfer of care, and transition completion (Got Transition, c2020).

The survey was constructed in such a way that the health care provider participants had the opportunity to relate their experiences when caring for the patients outside of their regular scope of practice (as in caring for an adult in the pediatric setting), express an opinion concerning when should transition start, identify some of the barriers that currently exists and propose resolutions to address these problems. Thus, the types of survey questions included: a) single answer questions which allowed respondents to choose only one option, b) dropdown questions which gave the participants a list of answers from which to choose one, c) demographic questions which provided the opportunity to get better understanding of the professionals’ responses (RN, MD, PA, NP) in different clinical areas, and d) open-ended questions which gave the participants the opportunity to identify some of the barriers to transition, strategies that could be implemented to address the issue, as well as provide suggestions regarding the structure and process of a transition program.

3.5.4.2. Data Management and Analysis

The Survey Monkey data set was divided into two categories: one where the closed-ended questions were tabulated and organized for easier descriptive analysis. The open-ended responses were organized and coded thematically. The main codes for this portion of the analysis were guided by the domains and elements from the Patient Centered Medical Home and National Quality Forum for Measuring Care coordination and were centered around the research questions. Themes from each group were
tabulated and compared to identify the common threads in understanding the problem.

The final part of the analysis involved the integration of the qualitative and quantitative findings for the purpose of identifying a common thread and for understanding why young adults are still receiving sickle cell disease care in pediatrics.
CHAPTER FOUR

RESEARCH STUDY RESULTS

PATIENT PARTICIPANTS (INDIVIDUAL INTERVIEWS)

4.1. Introduction

This chapter focuses on the results of the qualitative portion of the study, which involved individual interviews conducted with young adults aged 18 to 30 years old who had sickle cell disease. Ideally, during the transition process from pediatric to adult care skills are developed that promote autonomy, personal responsibility and self-reliance. This chapter reports the results of individual interviews of young adults with sickle cell disease because their insights and experiences are fundamental to identifying why young adults remain in pediatric care years after they should have transitioned to adult care.

The research questions considered in this chapter were designed to gather information about barriers to and facilitators of transition, as well as to elicit information regarding the current state of transition, participants’ experiences with transition and their perceptions of when transition should begin, as well as how it would be most beneficial. Therefore, the study addressed the following major research questions: 1. What are the individual, provider and system factors that patients perceive as facilitating and/or impeding transition from the pediatric health care system to the adult health care system for young adults with SCD? 2. Are younger adults (aged 18 to 30 years old) with SCD prepared to navigate the adult health care system? 3. What are health care providers’ perceptions of patient, system, and providers factors that impact transition? 4. What are health care providers’ and patients’ perceptions of how and when transition from pediatric to adult health care services should begin? This chapter presents the analysis
and findings of the concepts, codes and themes that address these questions, focusing on patient participants.

### 4.2. Description of the Sample

Demographic information regarding participants include gender, age, educational level, employment status, ethnicity/country of birth, and place of care (pediatric or adult health care service). Twenty-four participants signed consent forms in order to participate in the study: 10 females (42%) and 14 males (58%). Sixty-seven percent (16) of those who signed consent forms were interviewed. The interviews were conducted between April 2016 and November 2017.

The figure below depicts the number of participants who signed consent forms and who were interviewed.

![Figure 2: Participants who signed consent forms and were interviewed](image-url)
A total of 4 females and 12 males completed interviews. The researcher was unable to schedule interviews with 6 females and 2 males who signed consent forms. These participants ranged in age from 18 years to 30 years, which was the intended age range of the research study. The male participants ranged in age from 18 to 30 years while female participants ranged in age from 22 years of age to 25 years of age. The ages of the 8 participants who signed consent forms but were not interviewed were not captured because information regarding age was obtained at the time of the interview.

Eleven of the patient participants were born in the United States, 3 were born in Africa (each in a different country) and 2 were born in the Caribbean (each in a different country). The countries of birth were not reported to maintain participant anonymity. All participants (N=16) were of African descent and none were of mixed-race ethnicity.

<table>
<thead>
<tr>
<th>Gender</th>
<th>GED</th>
<th>HS graduate</th>
<th>In college</th>
<th>Some college</th>
<th>College graduate</th>
<th>Vocational</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male (12)</td>
<td>1</td>
<td>4</td>
<td>5</td>
<td>1</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Female (4)</td>
<td></td>
<td></td>
<td>2</td>
<td>1</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Total (16)</td>
<td>1</td>
<td>4</td>
<td>7</td>
<td>2</td>
<td>1</td>
<td>1</td>
</tr>
</tbody>
</table>

Table 5: Educational level of patient participants

The educational level for participants ranged from those who received their GED to college graduates. Table 5 depicts the educational level of these participants. One of the male participants was a full-time graduate student and was included in the “in college” group. The participant who graduated from college was employed full-time.
Seventy-five percent of the study population were employed; 12 individuals worked either full time or part-time and 4 were unemployed.

Figure 4 summarizes where participants receive sickle cell specialty care.
Male participants represented 75% of participants (N=12) while female participants represented 25% (N=4) of the study population. Two of the male participants received health care services from both pediatric and adult hematologists at the same institution. One participant reported that he continued to go to the pediatric clinic because “I don’t want to leave.” He had experienced receiving services in adult clinic and adult emergency department but did not want to be admitted to the adult hospital “because they don’t have privacy there.” When probed, he reported that he was once admitted to the adult hospital and he had to share a room with an elderly person. Two participants receive their care exclusively in adult health care services. One participant did not follow-up in either clinic; he only accessed health care services through the emergency department when he was ill. He was treated in the pediatric clinic when he was a child. Upon turning 21 years of age, whenever he accessed emergency health care services, he was treated in the adult emergency department. However, if admission to the hospital was warranted, he was admitted to the Children’s Hospital on the pediatric hematology service. He reported that after discharge from the hospital, he never followed up in the pediatric clinic because he “felt strange sitting around with little kids.”

Three of the female patients received regular health care services in the pediatric clinic even though they were all over the age of 21. The other participant received her care in the adult clinic. Of the 16 participants, 11 were 21 years old and older but only 3 were receiving health care services in the adult clinic. The other 2 participants were under the age of 21 years of age and received their care in pediatrics.
4.3. **Sample Summary**

Twenty-four individuals signed consent forms for the study and 16 individuals (67%) were interviewed. There were 12 males who ranged in age from 18-30 years of age and 4 females who ranged in age from 22-25 years of age. Sixty-nine percent of the participants had some education beyond high school with 7 (44%) currently in college; one had graduated from college and was in the process of applying to graduate school, and one individual had completed vocational school. Seventy-five percent were employed. Eleven (69%) of the participants were born in the US and 5 (31%) were born in another country (Africa and the Caribbean).

4.4. **Study Setting**

The pediatric hematology/oncology clinic is located within the Rutgers Cancer Institute of New Jersey. Pediatric patients with cancer and all hematology conditions (with the exception of hemophilia) are treated in this clinic. Although it is a pediatric clinic, the age of the patients ranged from infancy through 25 years of age. The adult hematology clinic is located in the Clinical Academic Building on the same campus. The latter is operated under Rutgers Health/Robert Wood Medical Group. Although both clinics are located on the same campus as Robert Wood Johnson Barnabas Health Hospital, New Brunswick, they are not in the same building.

There are two separate emergency rooms, one that serves patients from birth to 21 years of age and another that serves patients over the age of 21. Both emergency rooms, pediatric and adult, are located at Robert Wood Johnson Barnabas Health Hospital (RWJHB) New Brunswick campus. These emergency rooms are located close to each
other. The children’s hospital and the adult hospitals are free-standing hospitals with separate entrances.

4.5. **Results**

The interviews, which followed a semi-structured interview guide, were transcribed verbatim and the transcripts were imported into QRS NVivo 10 for organization and data management.

As a reminder, data analyses were carried out in three stages through direct content analysis using the constant comparative method (Boeije, 2002; Charmaz, 2014). The transcripts were read in an iterative process to identify themes. Similar themes were grouped together and placed under the concepts that best described them. Themes emerged from the data organically. All themes were then labelled and categorized based on each research question (Appendix 6). The data were further examined from the perspective of those young adults who were still in pediatric care and those who were in adult care in an attempt to identify similarities, differences and relationships in the answers to the research questions. As a result, the same text was used for different codes as long as the opinions of the participants and the context were similar. The categorization of the codes under each concept allowed for the identification of patterns that generated the themes. Descriptive statistics (percentages and frequencies) are used to describe demographic variables.

Five concepts generated from the research questions and a review of the transition and sickle cell literature formed the basis for the organization of the data. The key concepts are *transition, access to adult health care services, sickle cell disease,*
information technology and communication. A thematic framework that was developed around elements of the Patient Centered Medical Home model (Agency for Healthcare Research and Quality) and the National Quality Forum (Antonelli et al., 2009; National Quality Forum, c2021) framework for measuring care formed the template for organizing codes and guiding data classification. A list of codes (Appendix 7) developed from the literature review and categorized under each concept, was used to organize the data, thus allowing for the identification of patterns that generated themes. The constant comparative method (looking for similar codes across interviews) was used to generate themes relating to the research questions. Each theme was supported by quotations from the patient participants during the individual interviews. The data were concurrently collected and analyzed using an inductive, iterative approach, utilizing the precepts of grounded theory (Charmaz, 2006; Strauss & Corbin, 2007).

For clarity, each key concept is highlighted in bold letters. Transition is the process of moving from pediatric to adult health care services, exemplified by actions that ensure that there is coordination and continuity of health care services for young adults with sickle cell disease who age-out of pediatric care. As adolescents and young adults (age 18-30 years) transition to adult health care services, they encounter barriers that hinder them from effectively crossing the chasm between pediatric and adult health care services effectively. Codes that are categorized under the concept of transition include barriers to transition, facilitators of transition, timing of transition, preparation for transition and experience with transition.
4.5.1. **Barriers to transition**

Barriers to transition are those processes that prevent transition from occurring in a timely manner. Barriers to transition were divided into three categories: individual (patient-related), health care provider and system barriers. **Individual barriers** include but are not limited to lack of knowledge about transition, sickle cell disease and its complications and the importance of ensuring that there is ongoing clinical supervision. **Health care provider barriers** include but are not limited to lack of time to spend with patients, poor preparation (by pediatrics) for the adult health care system, poor communication between health care provider and patients or family members. **System barriers** include absence of established transition policies and protocols, inadequate numbers of health care providers and ancillary staff, inadequate educational information about sickle cell and transition, insufficient appointment openings to accommodate patients, and lack of appropriate community linkage to services that promote and maintain high quality health care. **Facilitators of transition** are those processes that enable young adults to effectively transfer care from pediatric to adult services. Having the people who are affected articulate what processes are beneficial in assisting them to effectively leave pediatrics is necessary to resolve the problem.

The other codes that were assigned to the concept of transition are as follows: **readiness to transition**, which is a feeling of preparedness that one can navigate the adult health care system; **preparation for transition**, which are the necessary steps that are needed to develop confidence to enter the adult health care system, and **timing of transition**, which stipulates the ideal time to start transition planning and describes the time when transition is over and when transfer should occur. **Experience with**
transition, which is the expression of the lived experiences of patients as they age-out of pediatric care, and how transition should be done were other codes included under the concept of transition.

**Individual (Patient-Related) Barriers to Transition**

These include the lack of knowledge about:
- Transition
- Sickle cell disease and its complications
- The importance of ensuring that there is ongoing clinical supervision

**Health Care Provider Barriers to Transition**

- Lack of time to spend with patients
- Poor preparation (by pediatrics) for the adult health care system
- Poor communication between health care provider and patients or family members

**System Barriers to Transition**

- Absence of established transition policies and protocols
- Inadequate numbers of health care providers and ancillary staff
- Inadequate educational information about sickle cell and transition
- Insufficient appointment openings to accommodate patients
- Lack of appropriate community linkage to services that promote and maintain high quality health care

**Other Variables Related to Transition**

- Readiness to transition
- Preparation for transition
- Timing of transition
- Experience with transition

Figure 5: Barriers and other variables related to transition
4.5.1.1. **Research Question 1 and Key Concepts: Barriers to Transition**

Transition is “the purposeful, planned movement of adolescents and young adults with chronic conditions [like sickle cell disease] from pediatric to adult care (Blum et al., 1993). Successful transition from pediatric to adult health care services for adolescents and young adults with sickle cell disease is a major challenge. In order to understand the challenges that these individuals encounter, this researcher sought to identify the factors that adolescents and young adults perceive to impede and/or facilitate transition from pediatric to adult health care services. **Transition** is a set of actions that ensure that there is coordination and continuity of health care services for young adults with sickle cell disease as they age-out of pediatric care. The ideal transition is comprehensive, well organized and occurs over time. The themes generated by the data are categorized and reported in three sections: individual (patient-related) barriers, health care provider barriers and system barriers.

4.5.1.1.1. **Individual Barriers**

There are multiple factors that were attributed to patients that prevented them from seeking or accessing health care services that were age-appropriate and necessary to promote and maintain health. They included behaviors that impacted the management of their chronic health condition, especially as they transitioned from pediatric to adult health care services. Knowing what transition meant to each of the participants was important because the meaning ascribed to the word may influence the decisions that are made to seek age-appropriate health care services. As such, each participant was asked what transition meant to them.
Study participants gave a range of definitions: A 21-year-old male described transition as,

“I just know that I will get a different doctor and all the information they had on me in pediatrics will be given to the adults. I am assuming that the care will be the same, but that’s really all I know. I don’t know anything about transitioning.”

Another 21-year-old male who only goes to the emergency room whenever he has pain, but never follows-up for routine sickle cell care, outrightly indicated that he was not familiar with transition. “I don’t know anything about transitioning from pediatric to adult. Just figured that whenever I turned twenty-one, I’d be sent to adult care.” When probed, he responded, “I have to be more responsible for my results, getting them myself and following up with them more instead of them following up with me.” He expressed that he was not prepared to go to adult care “because I am very undereducated about sickle cell.”

A 22-year-old college student expressed her opinion about transitioning to the adult system this way:

“I would be so upset! I would be so upset! I would be ready, but I would be so upset. I would be upset because, like I said, the convenience and the comfortability and the people that I know in Peds [pediatrics]. All my friends are there, and stuff like that, so it would just be the insignificance of moving on.”

This participant had no plans to ever leave the pediatric system because she was comfortable with her health care team and just could not envision going to new provider(s). She was not the only participant who was adamant that they were comfortable in the pediatric setting. Four other participants over the age of 21 who were still receiving their care in the pediatric setting articulated similar sentiments. A 23-year-old male said,
“Because I know everybody here and everybody here knows me and my history. And I feel comfortable with them. And they always go the extra mile to help me out. I like my doctor a lot, it’s a comfort thing. When I’m comfortable with something, I don’t want to change”.

Another participant said, “I am 22 already, and I’m very comfortable being in pediatrics. I would hate to be put into the adult world right now being uncomfortable with it, being unprepared, and just thrown into that.”

Those participants over the age of 21 years who were still receiving their health care services in pediatrics but who, when they were ill, received emergency health care services in the adult emergency room also expressed fear about leaving pediatrics. One participant shared his fear of going to the adult emergency department and his experience with his first exposure to the adult health care system this way:

“what really happened was one night, I got really sick. I came in an ambulance and I told the [emergency medical technician] EMT that my doctor was a pediatric doctor, so he took me to the pediatric emergency room. I was lucky because I don’t look my age. The next time I had a crisis, I couldn’t speak, I didn’t have the energy to speak and I didn’t have anyone to advocate for me, so they looked at my age and I was thrown into the adult world, alone.”

Although this participant was aware that at the age of 21 years, he would receive emergency sickle cell care in the adult emergency room, he expressed that he was fearful to leave the pediatric setting and felt “alone” when he ended up in the adult ER for the first time. The fear of leaving pediatrics was heightened when he had to wait for a long time in the adult emergency department to be evaluated and treated by a physician. Although he had been receiving both routine clinical and emergency health care services in the adult system for over five years, he expressed that it was with much hesitancy that he left the pediatric setting. So, even though he was aware that at age 21 he would be treated in the adult emergency department, he took no steps to become informed about
the adult setting. Instead, he hoped that his youthful appearance would be sufficient to get him treated by the pediatric team.

A 30-year-old male who was already receiving all his care in the adult service expressed anger about how he got to adult service. His advice to those who were still receiving care in pediatric was,

“just make sure you be up on that pediatric doctor. Listen, I'm not going to leave, or I don't want you to send me over without finding me a hematologist, talking to the hematologist, telling them everything.”

A 23-year-old male who was still receiving care in the pediatric clinic summed up his transition preparation like this, “I am probably prepared, but I don't want to leave pediatrics.” Despite being over 21 years of age, these participants displayed a level of complacency about transition partly as a result of the absence of a transition program, but mostly because of their fear of leaving the pediatric health care system. Those participants over the age of 21 years who received primary hematological care in pediatrics but received emergency health care services in the adult emergency department expressed heightened fears of leaving pediatrics because of their personal experiences. Instances such as long wait times to receive treatment, getting inadequate pain medication, and feeling “judged as a drug seeker,” were some of the reasons that engendered anxieties about leaving the pediatric setting. The lack of preparation of young adults transitioning to the adult system resulted in patients falling out of regular maintenance health care.

The core underlying meaning of transition that was elicited from participants was that the physical place where they got care (the clinic) would change, they would be assigned a different doctor and they would have to take on more responsibility for their
own care. Education, age and place of care (pediatric or adult service) made no difference in how these participants defined transition. Transition, as described by all these participants, was seen as an event rather than a process. However, in later discussion (and with active probing) participants who were asked about what an ideal transition program should include clearly outlined a gradual process that would be individualized and organized (this will be discussed in detail later in the conclusion chapter).

The physical movement from pediatrics to adult health care services was not the dominant focus. Eleven participants (69%) age 21 and older were still receiving sickle cell disease care in pediatrics. These study participants were comfortable in the pediatric setting and they trusted their pediatric health care providers. This, coupled with fear of isolation in the adult system, prevented them from getting age-appropriate sickle cell disease care. They perceived that pediatric care was gentler and kinder than adult care.

The following statement of a 30-year-old participant who stayed in the pediatric setting until he was 24 years old supports the finding that young adults were partly responsible for staying in pediatrics longer than they should. He summed up staying in pediatrics this way,

“... experiences in the pediatric world – peds [pediatrics] is like heaven to sicklers. As a sickler, when I was younger and I was in the peds [pediatric] world, it was easier to get seen and treated faster. They tend to contact your primary physician faster than the adult world, because in the adult world, they just want to get you better and out of the door as fast as they can because they have to cater to more people, so to speak. But in the pediatric world, that's the opposite. They try to take care of as many kids and save their lives.” [In pediatrics], “the nurses care more, whether male or female, and doctors care more.”

A 23-year-old participant who still received his care in the pediatric clinic but when ill got emergency care in the adult emergency department expressed fear of isolation in the
adult health care system and thus fear of leaving pediatrics. This was exemplified by his statement,

“The treatment that I saw being in the [adult] emergency room, once you are an adult, you are on your own. Nobody really cares how you feel about certain things. The nurses don't pay attention as much, they don't come or attend to you in like reasonable time. I feel the pediatric side is more helpful and attentive and kinder. But now it’s like nobody – you are on your own.”

Quotes from these participants revealed a major concern. Sickle cell disease care in the adult health care system engendered anxieties and fears in young adults. The fact that these participants perceived the adult system, especially the emergency department, as uncaring is problematic. The perception that health care providers did “not care” could result in delays in accessing emergency care. Any delay in accessing emergency care for an individual with sickle cell disease could lead to deleterious outcomes, especially for an already marginalized population.

The primary individual barriers experienced by these participants included lack of knowledge about transition, trust of their pediatric health care providers, comfort with the care that is provided in the pediatric setting (acute and routine), and fear of leaving pediatrics and assuming full responsibility for their own care, as well as isolation in the adult setting.

4.5.1.1.2. **Health Care Provider Barriers**

Pediatric health care providers are responsible for educating patients about their health conditions and preparing them to assume responsibility for their health care. It is critical for young patients to develop the necessary skills to navigate the adult health care system. This is especially critical for those who are cognitively impaired as a result of
strokes (Ohene-Frempong et al., 1998). Six participants reported that they were told that ‘someday’ they would no longer receive health care services in pediatrics. A 25-year-old female said it this way, “She [health care provider] explained that I’m getting older, and I have to see another doctor. She said it’s going to be different, but that’s all I know.”

Another young adult who was getting health care services in the adult clinic described her experience this way: “…. nobody prepared me for the adult side. You know, pediatric nurses and some doctors, they always used to talk to me about like, you’re soon going to go the adult side, but they never told me the exact way.” She started receiving health care services in the adult setting when she was 23 years old. A total of 10 participants over the age of 21 (N = 14) reported that at least one clinician (nurse or doctor in the pediatric setting) mentioned to them that they would leave the pediatric clinic, but they were not provided information on what to expect and there was no discussion about timelines for their transition or how transition to the adult system would occur.

Four participants had a different experience with their health care providers. They contended that transition to the adult system was not discussed at all. This is reflected in the following statements. A 20-year-old male said, “The only thing I know about adult care from pediatrics care – well, I haven’t really been told anything. The only thing I think was told that the TV isn’t free or something. I've heard that before. I don't really know much about adult care.” [The television service in the Children’s hospital is free]. A 30-year-old male described his experience with transition this way, “I don’t know about other patients, but with me, I felt the ball was dropped. No, nobody prepared me for the adult side. I was in pain … I just want to be treated but they told me to sit over
Anticipatory guidance (counseling that takes place at each health care provider encounter) addresses the physical, emotional, psychological and developmental changes that will occur over time. Anticipatory guidance is a crucial element of pediatric health care and health care providers have a responsibility to provide anticipatory guidance. This guidance starts in infancy with the parents and gradually shifts to the patient as they age (Syverson et al., 2016).

Anticipatory guidance for adolescents with chronic illness should include discussions about the patient’s health condition, health risks such as smoking, sexual behavior and contraception, reviewing complications of sickle cell disease, and more specifically, the patient’s health status and the complication(s) that are specific to that patient, as well as guidance on practicing healthy lifestyle habits and adherence to medication regimen. However, most participants (N = 10) reported only cursory mention of transition. More than 50% of participants over the age of 21 years expressed disappointment in the way they ended up in adult care. More than 75 percent of the participants over the age of 21 years who received care in the adult setting (emergency department or clinic) did not experience transition planning as part of anticipatory guidance by their pediatric health care provider.

Only one participant reported that he had a positive transition experience. He reported that he met the adult team at least once and went back to his pediatric hematologist before he transitioned fully to the adult service. He officially transferred care from pediatrics to the adult system when he was 24 years old. Although he did not
leave pediatric care by the age of 21, he was satisfied with the way his transition occurred. He also suggested that the discussion about transition should start at an earlier age and that a transition program should include what to expect and how to navigate the adult system.

Overall, pediatric health care providers provided a caring and supportive milieu for their patients, but they did not effectively prepare these patients for entering and navigating the adult health care system. Participants reported that transition was mentioned by their pediatric providers, but it was not discussed. Consequently, they never focused on transition as a necessary process, despite admitting that they knew they would ultimately end up in adult care. So, as young adults aged out of pediatric care, they entered the adult system feeling unprepared, fearful and sometimes angry about the way they were treated in the adult system compared to the pediatric setting. Eleven patients (69%) 21 years and older were still receiving routine sickle cell disease routine care in the pediatric system. Although their pediatric health care providers continued to provide health care services to them, anticipatory guidance about transition was limited to an occasional mention that someday they would go to adult care.

Participants reported that they lacked the necessary skills to confidently manage sickle cell disease in the adult setting, as well as lacking knowledge about how to competently navigate the adult health care system. Navigation of the adult system included scheduling or rescheduling appointments, renewing prescriptions, and effectively communicating with health care providers. One participant summed up his experience this way,

“For starters, they can inform me about the transitioning, what to expect and what not to expect just to make things smoother so I can have an understanding of
Pediatric providers’ willingness to continue providing care to adult patients who have aged-out of pediatric care while not preparing them for the adult system is a major provider barrier. Eleven participants who were 21 years and older, of whom three were receiving all their health care services in the adult system, stated that they were not prepared to transition. It is the responsibility of the pediatric team to prepare these young adults to find their way in an uncoordinated, oftentimes fragmented adult system. Pediatric providers should incorporate transition as an element of anticipatory guidance. However, in order for pediatric providers to overcome some of these barriers, a well-devised transition policy with dedicated staff to educate patients and coordinate and implement strategies is necessary to ensure effective transition from pediatric to adult health care system.

4.5.1.1.3. **System barriers**

System barriers are those administrative processes that prevent patients from participating in a coordinated care transition from the pediatric to the adult health care system. System barriers also prevent participants from acquiring the necessary skills to navigate the adult health care system competently. Statements categorized as system barriers were expressed by all participants who were already receiving their care in the adult system, whether in the emergency department or in the adult hematology clinic.

Six young adults who were still receiving care in pediatrics expressed hesitancy about transitioning to adult health care services. However, they were resigned to the need
to do so if they could be adequately prepared. One 23-year-old participant summed it up this way, “

‘what I think would be really helpful is, instead of waiting until someone’s twenty-one, twenty-two to start talking about transitioning into adult hematology care, I think it would be helpful to start talking about it, having these information sessions when they're fifteen and sixteen. Then when they're eighteen, bring in the transition program.’

This response was echoed by several participants. The health care system does not currently dedicate adequate staff and/or time to teaching young adults about sickle cell and the various complications that are associated with their condition.

Hearing the voices of those individuals who were already in adult care is important in identifying how they experienced movement from the pediatric to the adult care system. This sample included four participants who were receiving sickle cell care in the adult service. One participant described going back and forth between the pediatric and adult provider and three were receiving all their care in the adult system. The participant who went back and forth received health care services in the adult emergency department but did not follow-up in the adult outpatient clinic. Therefore, this analysis focused on the participants who were in consistent care in the adult system.

A 30-year-old participant described his transition with disappointment. The following is the description of the experience, “Well with me, I don't know about other patients, but with me, I felt like the ball was dropped. It wasn't a smooth transition. No, nobody prepared me for the adult side.” Probing questions were asked to give this participant the opportunity to elaborate. The following quote is a description of his experience:

“I was in pain, so it didn't really dawn on me too much. I just want[ed] to be treated but they told me to sit over there [pointed to where adults sit]. So, I went
to where the adults sit. That's how I knew I was going to be over on the adult side.”

The abrupt placement of this participant in the adult system resulted in his dropping out from routine clinical care due to inadequate preparation for transition.

A comprehensive departmental transition program could have remedied the situation. Another participant’s similar statement also supports the assumption that addressing the issue from a system perspective would help young adults cross the chasm from pediatric to adult health care more effectively.

“I went two years without a hematologist, and about a year without a primary doctor until I spoke to certain people on the adolescent side. They said at the time, the adult doctor wasn’t taking no more sickle cell patients. So, they wanted me to transfer to another city; and I’m like, I don’t know nothing about Newark and Newark don’t know me, and I live closer to Robert Wood. I would love a doctor around here. But the doctor [adult hematologist] at the time was full to capacity. So, for those years, my primary doctor acted like my hematologist, but was getting help [from other doctors]. I guess she was calling and getting help to treat the sickle cell, you know, speaking to a hematologist. After two years, that's when the adult doctor [adult hematologist] had gotten help, another doctor. I don’t want to say his name. But that's how I had gotten a hematologist, when I got this hematologist.”

One would assume that in a tertiary care academic facility there would be adequate staff to treat patients with sickle cell disease. But this participant’s experience highlights the shortage of staff to treat patients with sickle cell disease. This issue goes beyond the departmental level and rises to an administrative system level.

This experience was not unique. The inability to get an appointment with an adult hematologist was experienced by all three participants who reported receiving their care in the adult clinic. One of these participants expressed his frustration with the adult system in this way:
“When you are an adult, they want to see you every three months, but you get sick often. That's not fair, that's the part that bothers me. In pediatrics, I was getting blood transfusions every four weeks [because of history of stroke]. My adult doctor was saying, 'see me every three or four months, and when you get sick, go to the ER.' That time in my life, that's when I was really mad because I felt betrayed.”

Another twenty-five-year-old female participant reported a similar situation regarding access to health care providers in the adult system. “In pediatrics, they work five days a week, both morning, noon, and evening. My adult hematologist only works from 1 o’clock, she only sees sickle-cell patients every Monday in the afternoon.” Clearly, the issue of adequate staffing and inadequate time slots for seeing patients must be addressed at the system level. Overcoming these major barriers will enable young adults with sickle cell disease to continue to receive the same quality of care that they all reported that they received in the pediatric system. It would also prevent circumstances like the above from occurring. In the case of the participant who was receiving blood transfusion, there was no systematic pathway for him to continue this stroke-preventive treatment plan when he entered the adult system.

The major system barriers to transition were the lack of transition preparation in the pediatric system, the absence of a clear transition policy and the lack of a transition program that outlined in an organized way to effectively transition young adults with sickle cell disease to the adult system. The lack of a transition policy and a transition protocol resulted in a haphazard transition, with some participants expressing feelings of abandonment and betrayal by the pediatric provider. One participant summarized it this way,

“I know patients that were kept over until they were 25. So, I felt some type of way, you know, how they treated me. I felt a type of way because I still had friends that were over there, and they were of age. And now I'm over there on the adult
Difficulty accessing care services due to limited clinician availability to provide sickle cell disease care is also a major system barrier.

4.5.1.2. Facilitators of transition

Facilitators of transition are those processes that allow young adults with sickle cell disease to effectively transition from the pediatric to the adult health care system. Although the researcher elicited responses from all participants about what constituted effective transition, the analysis of facilitators of transition was limited to those individuals who were getting their care exclusively in the adult system. This decision was made because the question was more relatable for those who were already receiving health care services in the adult setting.

Three participants were able to identify facilitators to transition, although only one articulated that she had a positive experience during her transition. This twenty-five-year-old participant’s recollection of her perception of what helped to facilitate her transition was as follows: “My doctor told me that I am getting older and will have to see another doctor and she would refer me to someone really nice who would take care of me.” She reported that her pediatric hematologist made the initial appointment for her and she considered that a facilitating factor. When asked if she felt prepared for the adult system, she replied, “I wasn’t prepared enough because I did not know that their rules were different.”

When probed, she reported that there have always been times when she felt pain and was unable to go to school and work. When she was under the care of the pediatric
hematologist, whenever she was unwell, she was always able to speak to someone (a nurse or doctor) from the pediatric clinic, because the clinic was open all day Monday to Friday. She was also able to get an excuse note for her employer if her sickle cell crisis prevented her from going to work. However, since the adult clinic only operates one afternoon per week, there was no one to contact for such accommodations once she transitioned to adult care. She also reported that no one was willing to write an excuse for work unless the patient was first seen in the clinic. However, with limited office hours and booked clinic schedules, it was almost impossible to be seen outside of regularly scheduled appointments. Furthermore, when one is unwell, providers in the adult system automatically refer patients to the emergency department.

The other two participants were unable to recall any experience that they would describe as positive in facilitating transition to adult health care services. However, they were eager to make recommendations about strategies that could be implemented for effective transition. Such strategies included a comprehensive curriculum that included information about sickle cell, complications of sickle cell, health insurance, health care proxy, access to medication when patients experience painful sickle cell crisis without having to go to the emergency room, and what to expect in the adult health care system. One participant proposed a team approach that would include pediatric and adult health care providers, a counselor or social worker, the patient and a caregiver. These recommendations will be summarized and discussed in the last chapter of this dissertation.
4.5.2. **Access to Health Care Services**

The concept of access to health care services for adolescents and young adults with sickle cell disease refers to the ability to obtain preventative clinical and acute health care services in an age-appropriate setting. Access barriers, therefore, are defined as those factors that prevent or make it more difficult for individuals to effectively use health care services. Health care access includes access to health care providers who can competently treat sickle cell disease, access to health care facilities with specialists who can treat patients with sickle cell disease and access to a health care system that has the infrastructure to provide comprehensive, ongoing age-appropriate health care services.

Another important element of a health care system includes the ability to access social services, health insurance, and community resources like case management. The following sections present themes that emerged regarding access to care and health care systems. Themes are categorized under three main codes: pediatric care, adult care and experiences in both pediatric and adult care. They identify barriers and/or facilitators of transition that are specific to each health care setting. This aids in the identification of areas that can be targeted for improvement. It also provides guidance for the development of a blueprint for addressing those deficiencies that are hindering timely transition from the pediatric to the adult health care system.

As a reminder, all patient participants (N = 16) received sickle cell care in the pediatric clinic, either currently or at some time in the past. During the study period, 3 participants aged 25 years and two aged 30 years were receiving all their care in the adult setting (the hematology clinic and the emergency department), one 22 year-old participant used the adult emergency department when ill but did not follow-up for
routine care either in the pediatric or adult clinics, 5 participants (all over the age of 21 years) were treated exclusively in the adult emergency room for acute illnesses but got their routine sickle cell care in the pediatric clinic, one 22 year-old participant received acute illness care in the adult emergency department and had been admitted to both the adult and pediatric hospitals, and 6 participants (21 years and younger) received both acute and routine sickle cell care exclusively in pediatric settings (pediatric emergency department and pediatric hematology clinic). This is demonstrated in Figure 6.

![Number of Patient Participants and Place of Care](image)

**Figure 6: Number of Patient Participants and Source of Treatment**

4.5.2.1. **Pediatric Care**

There was universal agreement among all participants that access to pediatric clinical care, both for acute illness and routine care, was exceptional. As mentioned before, pediatric care in the emergency department continued up to the age of 21. Therefore, this analysis focused on those participants (N = 6) who reported that they were currently receiving all their health care services in the pediatric setting. All these
participants reported that it was easy to get an appointment with their hematologist, that they were satisfied with the wait time in the clinic, and that they were satisfied with the care provided in the pediatric emergency room and the children’s hospital when they were admitted.

In New Jersey, access to pediatric sickle cell disease specialty care and health care providers with the clinical expertise to provide services to these patients is limited to the 5 state-funded sites, some with satellite clinics, (described in the review of literature chapter) and affiliated hospitals that have dedicated pediatric hematology staff. Four participants had never been treated in a facility other than their usual place of care. Two participants had gone to emergency departments in hospitals that were closer to their homes, but after initial evaluation they were transported to the New Brunswick campus (usual place of care) for further management of their sickle cell disease related illnesses.

In the pediatric health care system, it is customary for the patients/caregiver to notify the pediatric hematologist on-call whenever the patient is experiencing acute symptoms of sickle cell (usually fever or pain). It is customary for the hematologist to then notify the health care facility prior to the arrival of the patient. The clinician who is providing emergency care communicates with the pediatric hematologist on-call after emergency treatment is administered to discuss whether the patient will be admitted to the hospital or discharged home. If admission is required, arrangements are made through the pediatric transfer center for the patient to be transferred to the Children’s Hospital in New Brunswick.

The following statement is illustrative of the process of accessing care in the pediatric health care system,
“even before you get to the emergency room, we had the option to contact whoever is on-call that night. The hematologist on-call that night, call them in advance and let them know that we are going to the ER.”

Thus, it was easy for this population of patients to access health care services both in the clinic and emergency department.

As is clear above, the pediatric clinic utilizes a family-centered, comprehensive team approach in the delivery of care. The team include nurses, a nutritionist and a counselor whose responsibilities include the education of parents, the referral and registration of pediatric patients in early intervention, assistance with health insurance, and making accommodations for the educational needs of pediatric patients based on the Section 504 of the Rehabilitation Act. This Federal legislation requires public schools to provide services and accommodations for eligible students with disabilities (US Department of Education, n.d.). Sickle cell disease is among the chronic health care conditions that fall within the scope of the legislation.

The pediatric clinic also has a full-time counselor whose scope of responsibilities includes liaison work conducted between patients and educational institutions to ensure accommodations such as extra time to complete examinations, allowing time for rest while in school, and ensuring that patients are not penalized for school absences as result of illness. The counselor is also responsible for providing sickle cell education to school personnel (teachers and school nurses) so that they understand the needs of the student and why accommodations are needed. Other responsibilities of the counselor include community referrals such as early intervention, summer camps and community enrichment programs. All study participants reported that they needed the services of the counselor and had positive appreciation for having her as a part of their health care team.
4.5.2.2. Adult Care

Participants reported various constraints regarding access to both emergency and routine preventive health care services. Three participants (ages 25 – 30) received all their care exclusively in the adult health care system. In terms of access to routine preventative care, all participants expressed that they had an average to difficult time scheduling an appointment in the clinic. The major barrier was that there was only one hematologist who provided care to patients with sickle cell disease. Although there were many hematologists in the department, most worked with patients who had malignant hematological disorders. Thus, a shortage of personnel is a major access barrier for young adults with sickle cell disease.

Study participants also reported that they had decreased access to providers due to limited clinic hours. Only one afternoon per week was allotted in which clinicians saw patients with sickle cell disease. The following is a statement from a 30-year-old participant who was asked about his ability to get an appointment in the adult clinic,

“It’s difficult, because in the adult world, it’s not like when I was in pediatrics when you could be seen in the clinic if you are sick. The hematologist now when you are an adult, they want to see you every three months, but you get sick often, and that’s not fair. That’s the part that bothers me.” His wife confirmed his statement by saying, “a lot of times when he calls for an appointment, they say if you are sick, you should go to the ER” [emergency room].

A 25-year-old college student also expressed concern about the clinic’s limited office hours. Her major issue with access was related to managing pain without going to the emergency department. Whenever she was ill,

“They usually refer you to the ER and I hate to go to the ER, especially the adult ER. Because in the adult emergency room, you have to wait a very long time. In the pediatric, you’re seen right away, and they check on you constantly, ask you are you still in pain, do you need more meds, or whatever. But in the adult, you
wait a long time, and they don't check on you as much. You have a very long wait if you do a testing.”

She reported that although she has a primary care physician, her primary care provider does not write prescriptions for narcotics for sickle cell pain. The adult hematologist did not want to write prescriptions for pain medicine either, so when she had sickle cell pain, she was told to go to the emergency department. She reported an instance in which she was so upset about that approach that she elicited the assistance of her pediatric hematologist and her primary care doctor to advocate with the adult hematologist on her behalf. Although she expressed disappointment that she had to sign a pain contract with the adult hematologist (the contract stipulated that she would not see any other doctor for pain medication), it alleviated the anxiety of having to go to the emergency department each time she had pain.

Other study participants shared similar experiences of stigmatization around prescription of narcotics for management of sickle cell pain. Pain is the most common cause of emergency department visits and hospitalization for individuals with sickle cell disease (Ballas & Lusardi, 2005; Brousseau et al., 2010b; Bou-Maroun, Hanba, Campbell & Yanik, 2018). Painful episodes are unpredictable and vary in frequency, intensity and duration and can be debilitating (Ballas et al., 2012). A study done by Payne and colleagues (2018) revealed that early administration of narcotic pain medication improved hospital outcomes. Despite knowledge that sickle cell disease-related painful episodes could be deleterious to the patients’ health, participants who received sickle cell disease care in both the outpatient clinic and emergency department struggled to get treatment in a timely manner. Other participants experienced the stigmatization that they were drug seekers, prompting one patient to make the following remarks: “Plenty of
times, believe me, you go into the ER and some doctors and some nurses, they think you are there just to get pain medicine. They judge you without knowing you.”

The other 30-year-old participant described his access to care as follows,

“Peds [pediatrics] is like heaven to sicklers [individuals with sickle cell disease]. It’s easier to get seen and treated faster. They usually have a protocol to follow, they tend to contact your primary faster than the adult world, because in the adult world, they just want to get you better and out the door as fast as they can.” If you are admitted, “they don’t contact the hematologist, they just call the house physician [hospitalist who only admit patients to the hospital but has no outpatient follow-up or contact] desk on call that night to come and admit you, or the admitting team. Whoever it is, they don’t even know your history. You are at the mercy of the general physicians who think that they know how to treat sickle cell disease, which is hard.”

These participants attributed the decreased access to adult providers in the clinic to limited operational hours (one afternoon weekly). This resulted in a reduced ability to get appointments, especially for sick visits.

Access to a hematologist was also limited because there was only one hematologist who provided care to patients with sickle cell disease. As previously stated, limited operational hours and decreased access to medical staff resulted in patients being referred to the emergency department for medical care. Study participants reported that sometimes they think their complaints could be managed in the outpatient setting.

When patients with sickle cell disease are admitted to the hospital, they are admitted under the care of a general hospitalist service and not under the care of a hematologist. Their medical care during hospitalization is managed by a doctor with whom the patients do not have a relationship. In these circumstances, the hematologist is simply a consultant. The participants reported that they would prefer that their care be directed by the hematologist, a doctor who knows them, whom they trust, and who is a specialist in sickle cell disease.
Access to emergency care was dependent on the hospital that they visited when they were experiencing fever or pain from a sickle cell crisis. All three participants reported that they have received care for acute illness in emergency departments at different hospitals that were in close proximity to where they were located when they started to experience symptoms. All reported that they had to wait for a long time to receive treatment and that they perceived that the clinicians were not adept at treating patients with sickle cell disease. One participant reported that she only goes to the hospital close to where she lives if she is unable to get a family member to drive her to Robert Wood Johnson Barnabas Health in New Brunswick.

She described her experience in the local emergency department as follows:

“the care was awful. You check in and wait like an hour or two before anyone sees you. Then a nurse comes, did my IV (catheter inserted into a vein) and took blood. Then I waited another hour before a doctor saw me and asked me a lot of questions. Then I had to wait another hour before I was able to get pain medicine. The medicine they gave me was Toradol, which only worked for ten minutes and the pain only got worse. She said that this has happened several times. She continued, “Although it is hard, most times, I would rather stay home than go there because I know the treatment that I am going to get. So, it’s like, why am I going to bother?”

When individuals like this participant experience sickle cell crisis, the emergency department is the only option they have to seek relief from the unrelenting pain that occurs. The delay in receiving timely evaluation and treatment with the right medication is partially a result of suspicion among health care providers that patients with sickle cell disease seek narcotics by faking their pain. This 22-year-old participant clearly articulated that sentiment: “Plenty of times, believe me, you go into the ER and some doctors and some nurses, they think you are there just to get pain medicine. They judge you without knowing you.” The discrimination and stigmatization that these patients
experience forces them to delay going to the hospital when they are experiencing pain. As mentioned earlier, vaso-occlusive crisis (pain) oftentimes triggers some of the most devastating complications of sickle cell disease (Ballas et al., 2010).

In an attempt to address the delayed and suboptimal management of sickle cell disease pain in the emergency department, the 2014 National Heart, Lung, and Blood Institute (NHLBI) Expert Panel Report on the Management of Sickle Cell Disease recommended that patients presenting to the emergency department with pain should be evaluated promptly and should be given pain medication within 30 minutes of initial assessment. Pain medication should include opioids and patients should be re-assessed every 30 minutes and additional medication administered until pain is at a level that can be tolerated by the patient (Yawn et al., 2014). The findings of my study suggest that health care providers in adult emergency departments fail to deliver the basic standard of care to patients with sickle cell disease despite clear evidenced-based guidelines from an expert panel.

The other two participants who received all their sickle cell disease care in the adult system articulated similar experiences when they described accessing health care services in emergency departments that were not at the study site. These findings suggest that although young adults with sickle cell disease have access to routine and emergency care, they face challenges getting timely, specialized care. Delay in receiving prompt, high-quality sickle cell disease care ultimately results in a range of negative health consequences to multiple organs (as indicated in review of literature) as well as significant emotional trauma.
4.5.2.3. **Experience in Pediatric and Adult Health Care Systems**

Five participants straddled both pediatric and adult health care systems. These participants, ages 21 – 24 years, continued to receive preventative health care services in the pediatric system; however, they received treatment for acute illnesses in the adult emergency department. Their experiences with access to routine health services were similar to those participants in the pediatric setting. It was easy to get appointments in the clinic and whenever they were ill, their pediatric sickle cell disease health care providers were accessible 24 hours/7 days per week. They were satisfied with the care that they received and the time that they spent in the clinic. They also trusted their pediatric hematologist and the clinic staff, but their experiences with access to health care services for acute illnesses in the adult emergency department mirrored those of the participants who were treated in the adult emergency department, as detailed above. A 22-year-old participant in this group described his experience in the adult care as

> “a lot slower and more uncomfortable, a bit more frustrating because it is difficult to get my point across. Instead of getting my medicine administered within 30 minutes, I would wait an hour or more to get my medicine administered. What I would like is for everything to pretty much be the same [as in the pediatric setting], so everybody is treated the same and nobody is ignored.”

The yearning for equal treatment in the adult emergency system as that which is standard in pediatrics was espoused by all participants who received care in the adult system. The difference in the delivery approach led this participant to draw this conclusion, “adults need attention too, pain is pain. If I have pain when I’m three, it’s the same as having pain when I am 30.”
4.5.2.4. **Summary**

Access to health care services, both acute and routine care, was easier in both the pediatric clinic and emergency department than in the adult system. The pediatric clinic was open on weekdays, giving patients the ability to speak with clinicians if they were experiencing any symptoms that can impact their health. Longer hours of operation also provided easier access for appointments and outpatient sick visits. There was always a physician on-call who returned all calls to the patient/caregiver after the clinic was closed. When the patient was referred to the emergency department, the pediatric hematologist on-call and the emergency department clinician discussed the patients’ clinical condition and the treatment plan prior to admission to the hospital or discharge home. When patients with sickle cell disease required admission to the hospital, they were admitted to the pediatric hematologist service. However, when they were admitted to the hospital in the adult system, the hematologist was not the clinician in charge of the treatment plan while in the hospital; he or she was only a consultant.

There were stark differences in the way care was organized and delivered in the adult system. There was only one hematology clinician assigned to care for patients with sickle cell disease and the clinic was open just one afternoon each week. This resulted in patients having minimal or no access to their hematologist on an emergency basis. Often, they were referred to the emergency department for any symptoms or illnesses that they experienced between their scheduled visits. Study participants who received health care services in the pediatric emergency department prior to ‘aging out’ reported that there was a vast difference in the way care was delivered in the adult and pediatric emergency
departments. This was captured in the following statement made by a 22-year-old participant,

“... the care in pediatrics is impeccable. I can’t explain it, I ask other sickle cell patients and we all agree when we say that the pediatric care at Robert Wood Johnson is incredible, that like we get the best care here.”

Another participant described the difference in care in these words:

“On the pediatric side, they’re always checking on you, making sure that you are walking, eating and taking your medication. So, I feel like they care more. And on the adult side, they treat you like an adult, like you are grown, you are on your own. They still check on you, but it’s not as consistent as it is in pediatrics.”

All participants who received their health care services exclusively in the adult setting, described their experiences in the emergency department as involving waiting longer to be evaluated by a clinician and even longer to receive treatment for their complaints. Experiencing a sickle cell pain crisis was the most frequent reason for accessing emergency services. It is important to note that sickle cell pain crisis cause tissue and organ damage and is often a prelude to a life-threatening condition like acute chest syndrome (Jain et al., 2017; Ballas et al., 2010) so prompt and aggressive treatment of acute pain is important in reducing complications and creating better health outcomes (Ballas et al., 2012; Yawn et al., 2014). Therefore, rapid evaluation and treatment of patients with sickle cell who presents to the emergency department is crucial.

4.5.3. **Knowledge of Sickle Cell Disease**

Knowledge about sickle cell is an important variable that may influence health care decisions such as keeping appointments for routine care, accessing care in a timely manner when they are experiencing pain and ensuring that there is no gap in services as they age-out of pediatric care. Data gathered to assess knowledge focused on questions
about knowledge of their own variant of sickle cell, the ability to name at least three complications of sickle cell disease and an awareness of the importance of receiving uninterrupted sickle cell disease care.

Eleven participants (69%) were not able to confidently describe the sickle cell variant that they carry. It is important to know the variant of sickle cell disease that individuals carry because some complications are dependent on the variant of sickle cell disease. For example, individuals with Hgb SS and S-β₀ experience more hemolysis and are at risk of complications such as avascular necrosis of the joints. But individuals Hgb SC and S-B+, which affects the small blood vessels in the body, puts individuals at risk for conditions like leg ulcers and renal failure (Ballas et al., 2010).

In this sample, level of education was not related to knowing the variant of sickle cell disease. Five participants, ranging in age from 21 to 22 years and with varying levels of education were not able to articulate what variant of sickle cell they carry. Two college students’ responses were, “I don’t know,” a high school student said, “I am not sure,” and another college student said, “the more severe one.” When probed, he was not able to name which variant was more severe. These participants’ lack of knowledge about their sickle cell variant was due primarily to lack of acquisition of sufficient information from their health care provider over the years in the pediatric system.

Nine participants (56%) were able to name at least three complications of sickle cell disease. Three participants were unable to identify any complications, despite living with more than one complication. A 21-year-old stated, “I don’t know, other than having to be in the hospital whenever it’s cold and when you get stressed, then that would be the extent of what I know [about complications]. A 23-year-old college student’s response to
naming three complications of sickle cell disease was, “I just know they have weak bone; I don’t know complications due to sickle cell that happen.” Four participants (25%) were able to state one or two complications of sickle cell disease as evidenced by the following responses, “I do have shortness of breath when I do activities like exercise, but other than that, I don’t know nothing else really.” Knowledge of the complications of sickle cell disease may portend better health outcomes. The participants who were not knowledgeable were receptive to gaining new knowledge.

Thirteen participants (81%) reported that they kept their follow-up appointments in clinic as scheduled. Four of thirteen participants were seen at least every 4 to 6 weeks because they were receiving chronic transfusion therapy. Three participants were not consistently receiving routine preventive care; 1only received episodic care in the emergency department and when he was admitted to the hospital.

The following are the responses of three of the individuals who did not have consistent follow-up care. A 23-year-old stated that, “I am supposed to see the doctor once a month, but that doesn’t always happen.” When probed, this individual was last seen four months prior to the interview and he did not have a scheduled appointment for a follow-up. When a 22-year-old male was asked how often he is seen in clinic, his response was, “never.” When probed, he was not able to articulate why he never follows-up. The participants not receiving routine preventive health care services were also among those who did not know the variant of sickle cell disease they carried and were not able to list the complications of sickle cell disease. This is important to note because an in-depth understanding of sickle cell disease and its complications may the catalyst for
empowering patients to become involved in preventive care by keeping their clinic appointments.

4.5.4. Communication

Effective communication between primary care providers, patients and specialty care providers is important for achieving successful coordination of health care and to prevent duplication of services. It is also important for the coordination of transition services because some insurance plans dictate that the participant must have a primary care physician. Effective communication also includes the patients’ ability to communicate with health care providers and vice versa as they build trusting relationships to support their overall wellness. In this study, communication was divided into three major categories: 1) Patient-to-provider communication, 2) provider-to-patient communication and 3) provider-to-provider communication in the pediatric and adult health care systems.

Study participants in the pediatric health care system reported that they were all comfortable communicating with their pediatric providers, including the nurses and other staff members. They had no inhibitions about discussing personal issues with their health care providers. They also expressed that their hematologists communicated effectively with them. Although communication about clinical care was effective, the patients’ deficiency in knowledge about sickle cell disease, its complications and their lack of preparation for transition suggests that communication about these areas was limited in scope. Seven participants who received care in pediatrics had primary care physicians and 5 received all routine care from their pediatric hematologist. Medical updates were
sent to primary care physicians after each visit clinic visit. When patients from the pediatric clinic access emergency health care services, their pediatric hematologists contacted the emergency department clinicians in advance so that when they arrived at the emergency department, the staff was already aware of their chief complaints. This communication shortened the time for the implementation of a treatment plan such as inserting an intravenous catheter, drawing blood and administering pain medication or antibiotics if needed and intravenous fluid.

The majority of the patients who had experience in the adult health care system expressed some degree of frustration with their communication with clinical staff. One 30-year-old participant said,

“You know, no matter how much you speak up for yourself, it depends on the doctor who is working. Because sometimes they feel as if when you go to the hospital and you are explaining to them what works for you. Some of them will have an attitude instead of thinking that you are trying to help them treat you. So, as I said, it depends on the doctor.”

All study participants who received care in the adult emergency departments at various facilities voiced similar comments.

They perceived that emergency department clinicians (physicians and nurses) dismiss their attempts to provide information about their health condition such as what medication works best for their pain. A 22-year-old participant relayed his experience this way:

“You know, no matter how much you can speak up for yourself, like I said, it depends on the doctor who is working. Because sometimes they feel as if when you go into a hospital, and you're explaining to them how they should treat you. It's like they don't like it because they think you are bossing them around, which is not really the point. You know what works and you are trying to help them treat you. So, some of them will have an attitude, some of them will not. So, as I said, it depends on the doctor.”
Effective communication in health care is bidirectional: both patient and clinician must understand each other. The interactions articulated by these participants suggests that patients with sickle cell disease have the added burden of proving that they are deserving of pain management when they are in pain crisis. Each participant was asked about how they overcome this challenge. The responses ranged from making sure that when they go to the hospital, they are accompanied by a family member who can advocate for them to “Sometimes, I just shut down and bear the pain.” These findings suggest that ineffective communication may result in the patient receiving suboptimal health care services.

The wife of a 30-year-old participant also supported the premise that patient-to-provider communication (and vice versa) in the emergency department has room for improvement. She said, “Because 90 percent of the time he’s here with crisis ... he has to fight with the doctors to take care of him. It’s like going to war when you are trying to get better because they make you feel like a burden.” Effective communication is important, especially in dealing with patients whose medical condition can be life threatening. From the clinician perspective, he/she need the proper clinical information to make the proper diagnosis and establish a treatment plan. From the patient’s perspective, he/she has to be able to articulate the symptoms and also feel heard and understood (Ong et al., 1995). In order to resolve this discordance, both patients and providers have to confront their biases and develop strategies for effective provider-patient (vice versa) communication.

Two of the adult patients in this study reported having primary care providers. One stated that his primary care physician and his hematologists communicated on a regular basis and the other stated that she did not know if her primary care provider
communicated with her hematologist. As indicated in this earlier part of the chapter, when a patient with sickle cell disease is admitted to the hospital, he or she is admitted to the hospitalist service, so the onus for management of the care falls on a clinician that the patient has no relationship with. A number of circumstances, as those indicated above, converge to cause a negative communication loop for patients with sickle cell disease in the adult system. However, the circumstance that has the most significant impact on effective communication is the availability of adequate staff in the adult emergency department and clinic, and more clinical hours in the adult clinic. Patients end up using the emergency department more frequently in their absence and some clinicians become impatient with patients who use the emergency department frequently. Patients tend to have longer wait times to be treated in emergency departments and the clinic, and when a patient is in pain for a long time without treatment, they become impatient with the clinician. This results in a negative feedback communication pattern in which both parties (patient and health care provider) can see each other as adversaries instead of allies working together to resolve the symptoms of sickle cell disease that is causing stress to the patient.

4.5.5. **Information Technology**

In this study, information technology included the use of computers, cellular phones and tablets in the management of health care for young adults with sickle cell disease. The ownership of a cellular phone, a tablet or a computer has the potential to improve patient outcomes and patient safety by improving medication adherence
(alarms), documenting health status, including medical and surgical history, and accessing health information through the medical portal.

All study participants (N = 16) owned a smart phone and 10 owned a tablet or a computer. All the patients who were receiving care in the pediatric clinic reported that they scheduled their follow-up appointments prior to leaving each clinic visit. Only two participants reported that they called to reschedule if they were unable to keep their scheduled appointment. Two participants used their phones to set reminders for taking medications, five used the calendar to keep track of their appointments and one participant used a sickle cell disease application to manage pain and anxiety. Seven participants participated in sickle cell disease on-line social groups. Three participants were aware that there was a medical portal where they could view their medical records and laboratory results, but only one participant had ever used it.

Although information technology devices have the potential to play a major role in the management of sickle cell disease, the majority of these participants were not taking advantage of the full scope of available technology. There are a number of applications for smart phones that they could use to manage their health. Some patients were unable to recall the number of times that they were admitted to the hospital, when they last had a blood transfusion and what medication works best for them when they were in crisis. Mobile telephones have the ability to interface with applications (apps) for documenting medical history, including transfusions, allergies and medications. There are apps that can be used for pain management and with the recent COVID-19 pandemic, these can be used for telemedicine to interface with health care providers.
Adolescents and young adults with sickle cell disease face a number of challenges as they age-out of pediatric care. The challenges are multifactorial and as a result, treatment requires a comprehensive approach with stakeholders from patient populations both in pediatric and adult hematology, clinicians from pediatric and adult hematology, emergency department, hospital administration and the New Jersey Department of Health and Senior Services (NJDHSS).

4.5.6. **Summary of Findings**

Study participants identified several reasons why it is difficult to leave the pediatric health care system. In particular, those who left, did so with much hesitancy and did so long after the time (age 21 years) when they should have done so. Participants’ trust of their pediatric provider is the major reason they identified for remaining in the pediatric system. This was partially a result of long-established relationships, some from infancy, and hearing stories that health care in the adult system is “awful.” A related theme that also emerged was fear. For those who are still in the pediatric system, there is fear of going to the adult system and dealing with clinicians “they do not know, and who do not know them.” For those who are receiving care in the adult system, fear was experienced differently and was focused on the care they received in the adult emergency department. They delayed seeking care in the emergency room because they feared being ignored and treated with indifference. A related theme that also emerged was that of been stereotyped as “a drug seeking” at a time when they were most vulnerable. In combination, these factors resulted in patients staying home and presenting to the
emergency department sicker than they would have if they had not been stereotyped and had gotten treatment in a timely manner.

Participants had **limited knowledge about sickle cell disease and its complications**. Education level did not make any difference in knowledge. Those participants who had less knowledge about sickle cell were less consistent with getting routine preventive care.

There is **minimal or no preparation for transition** for patients before they end up in the adult system. There are no established curricula, protocols or guidelines to aid or guide this process. Study participants saw transition as a process but reported that some providers only mentioned that “someday” they would go to the adult system and that was where the conversation ended. As a result, they lacked the skills to competently navigate the adult system.

At the administrative level (system barrier), the **absence of transition policy, limited clinical hours and staff in the adult clinic** were major themes that emerged. Using some of the elements of the PCMH model and NQF framework as well as incorporating recommendations from the participants, a transition model will be proposed in the conclusion to help to resolve the problem.
CHAPTER FIVE

HEALTH CARE PROVIDER PARTICIPANT SURVEYS

5.1. Introduction

This chapter provides the findings with respect to the following research questions: What are health care providers’ perceptions of patient, system and provider factors that facilitate and/or hinder the transition from pediatric to adult health care services? Are young adults with SCD being prepared to access and navigate the adult health care system? How and at what age should transition from pediatric to adult services occur?

As a reminder, children with sickle cell disease are surviving into adulthood. However, results from the recent Dallas Newborn Cohort revealed that majority of deaths in that cohort occurred within two years of transitioning to adult services (Quinn et al., 2010). McLaughlin and Ballas (2016) also reported that all the patients with sickle cell disease (eight out of 22) who were on chronic transfusion therapy because of history of stroke died within one to five years after transferring to adult care. They attributed this mortality to the discontinuation of transfusion therapy. This implies that failure to effectively coordinate the transition of this high-risk population will result in increased morbidity and mortality. In order to overcome this challenge, it is imperative to elicit the opinions of the health care providers who provide care to young adults with SCD. Their input is critical in identifying the reasons why young adults remain in the pediatric system longer than they should.

The primary goals of this inquiry are to explore health care providers’ perceptions of transition and how it should be done, as well as examine the processes of the current state of care for young adults with sickle cell disease to identify factors that contribute to
patients remaining in the pediatric system longer than they should. It is within this context that questions were developed to capture key insights from the health care professionals who provide health care services to this population.

The outline of the research was guided by elements in the Patient Centered Medical Home model (Jackson et al., 2013) and the National Quality Forum for measuring care coordination (Antonelli et al., 2009) as described in Chapter 4. This chapter follows a similar outline as the patient participant chapter. Five concepts generated from the research questions and a review of the transition and sickle cell literature formed the basis for the organization of the data. The key concepts are transition, access to adult health care services, sickle cell disease, information technology and communication. A thematic framework was developed following the precepts of grounded theory, whereby themes are identified from the text and categorized under the concepts. The constant comparative method (looking for similar answers in the surveys, linking them together and comparing their relationship) was used to build themes (Strauss & Corbin, 2007).

Since this study is exploratory in nature, the structure of the survey allowed health care providers to answer both forced responses and open-ended questions. There were fourteen forced- response questions and they were organized as single answer and multiple-choice answer selections, allowing respondents to select the response that best described their opinions. There were five open-ended questions, requiring participants to write-in the answers, thereby allowing them to express their personal perceptions about of why young adults are still in pediatrics and making suggestions as to how to resolve the problem. This approach allowed the researcher to get more in-depth information
from health care providers who work with young adults with SCD (pediatric hematology, pediatric emergency department, adult hematology and the adult emergency department).

Using the previously identified framework as a template, responses to the survey questions were categorized and coded around the elements of the Patient Centered Medical Home model (Mathematica Policy Research, Princeton, NJ, 2013) and the National Quality Forum for measuring care: Care Coordination (National Quality Forum, c2021). The data were further labelled and organized under specific research questions. Similar themes were grouped together and placed under the concepts that described them. Some themes emerged organically from the data. Themes were labelled and categorized based on the research questions.

As stated before, patients with sickle cell disease received their routine preventive and health maintenance care services from hematologists (pediatric and adult). However, individuals who are experiencing a sickle cell pain crisis, fever or any other sickle cell disease-related health complaints, access health care services through the emergency department. As a result, the survey was conducted among health care practitioners (RNs, NPs and MDs/DOs) in the pediatric hematology clinic, the pediatric ED, the adult hematology clinic and the adult emergency department. Physician assistants provide clinical care to patients in the adult emergency department and were included on the listserv of providers. However, none completed the survey, so they were not included in the analysis. No physician from the adult hematology clinic completed the survey.
5.2. Description of Sample

Demographic information regarding health care providers included gender, profession and area of employment. Thirty-eight participants completed the survey: 32 females (84%) and 6 males (16%). The web-based electronic survey (Survey Monkey) was used to collect data from health care providers (RNs, NPs, physicians) in the pediatric hematology clinic, the pediatric emergency department, the adult hematology clinic and the adult emergency department. The surveys were completed between March 28, 2017 and October 17, 2017.

The data collected from physicians in the ED did not identify whether the physicians worked primarily in the pediatric ED or the adult ED. Twenty-two registered nurses completed the survey (58 percent of the study population).

The pediatric emergency department nurses represented 59 percent of the nursing population. The adult emergency department has a higher number of nursing employees but only two nurses (9 percent of the study population) completed the survey. The adult hematology clinic is staffed by two nurses and one completed the survey. The results are summarized by place of employment and profession in each department as noted in the table below. However, in order to maintain anonymity based on place of employment, the results are reported by professional categories (nurses and physicians).
Figure 7: Summary of place of employment and professional in each department

5.3. **Research Question 3 and Results**

5.3.1. **Research Question 3**

What are health care providers’ perceptions of patient, system and providers’ factors that impact transition from pediatric to adult health care system for young adults with sickle cell disease?

5.3.2. **Results**

One open-ended question (question 14) was used to elicit direct responses to identify barriers to transition. However, answers to several forced response and multiple-choice questions (Appendix 5) also provided supplemental data that identified some of the contributory factors to the problem.

Since registered nurses spend more time with the patients during medical encounters, their responses were examined separately from other professionals (physicians and nurse practitioners) with the intent of teasing out any nuances that might
exist. Twenty-two nurses completed the survey. Five identified system and provider barriers but no patient barriers, and one participant did not answer the question. The diagram below illustrates the patient-related barriers to transition as perceived by registered nurses.

**Figure 8: Patient Barriers Reported by Registered Nurses**

Registered nurses across all departments reported that patients’ unwillingness to leave the pediatric setting was the most frequently identified patient-related barrier to transition. A nurse in the pediatric hematology clinic wrote “*patient does not want to transition to a new environment.*” Several nurses in the pediatric emergency department wrote statements such as “*unwillingness to transition, does not want to leave pediatrics,* *does not want to change doctors*” as patient barriers to transition. Young adults are more comfortable in the pediatric environment and as a result, they are staying longer in the pediatric setting.

Twenty-three percent of the respondents indicated that attachment or trust of pediatric health care provider is a major patient barrier to transitioning. In chronic health
conditions like sickle cell disease, patients tend to be cared for by the same hematologist for most of their lives. Consequently, they develop a bond that they are afraid to break. Nurses from different departments list the following as barriers to transition, “comfortable with their pediatrician”, “patients are comfortable with their providers,” and that “patients are attached to their pediatricians.” Patient participants also reported the trusting relationships with their pediatric health care providers as a major reason for staying as long as possible in pediatric care. They also reported distrust that a new provider in the adult health care system would understand their needs.

Twenty-four percent of the nursing respondents indicated that patients’ anxiety and fear of the unknown are barriers to transition. Anxiety and fear of the unknown barriers are identified exclusively by nurses who work in the emergency department (pediatric and adult). Quotations from the survey include, “fear of the unknown, nervous to leave the comforts of pediatrics, fear of leaving” [pediatrics], and “scared to leave” [pediatrics].

A small number of nurses cited issues such as, [patient] “doesn’t know how to take the first step”, “perception of greater access [to care]in pediatrics,” and “pediatric staff is more nurturing” as contributing to the reasons that young adults continue to receive their care in pediatrics. Although there were only a few such responses, they were included in the analysis because they are relevant; a significant number of patient participants expressed similar sentiments.

Four nurse practitioners and twelve physicians completed the survey. These health care providers (nurse practitioners and physicians) are responsible for evaluating and developing medical treatment plans for patients in regular health maintenance and
during acute illnesses. There was no way to differentiate the responses of those physicians who work in pediatric emergency room from those who work in the adult emergency department. No health care provider from the adult outpatient clinic completed the survey. The diagram below depicts the perceived patient barriers to transition as reported by health care providers.

**Figure 9: Health care providers’ perceived barriers to transition**

The 5 physicians in the pediatric hematology/oncology clinic did not identify any patient-level barriers to transition. Parents’ inability to give up medical decision-making and patient and caregiver reluctance to leave the pediatric health care system were the most frequently identified patient barriers to transition. Several emergency department physicians indicated that patients were not able to take responsibility for their care and that they behaved like younger children.

Patient/parent unwillingness to leave the pediatric setting was expressed by 54% of the health care clinicians. Health care providers also cited level of maturity as a contributory factor for young adults remaining in pediatrics. Level of maturity is the
patient’s ability to use their own health knowledge to communicate effectively with the health care provider. Patients with an acceptable level of maturity are able to take responsibility for the management of their own health. For example, they know what medications they are taking, they independently make appointments, and they are able to advocate for themselves, especially in the emergency situations. Statements such as, “not taught to take responsibility for themselves”, “lack of knowledge about own needs and how to make appointment,” and “maturity in self-care choices” are among the responses that health care providers cited and were categorized as patient barriers to transition.

Although data collection did not provide the opportunity for emergency department physicians to identify whether or not they worked in pediatrics or in the adult system, the responses provided by some participants suggested that they worked in the adult emergency department. Citations such as “parents hover and control everything,” and “perception that they are still kids” points to the challenges these clinicians may face especially as it relates to HIPAA and patient confidentiality issues for those patients over the age of 18. As children age into adulthood, some parents find it very difficult to relinquish control of medical decision-making, and as such may feel the need to be a little more protective of the young adult. This, however, can be viewed as a hindrance to the delivery of care in the adult health care system. Emergency department physicians have expectations that young adults should be more responsible for their own care and parents should be less involved.
5.3.2.1. **System and provider barriers**

These are combined because there are some factors that can be categorized as both. System barriers are those factors that negatively impact the efficient delivery of health care services. These issues are present at the operational and administrative level and if left unaddressed, can widen the health disparity gap. Provider barriers are those factors that impact the delivery of high quality, clinically and culturally competent health care services. Patients with sickle cell disease are already at increased risk for poor health outcomes, so identifying and addressing these barriers have important health consequences.

Ideally, transition from pediatric to adult health care services requires some degree of coordination between health care providers from both the pediatric and adult health care systems. The shared responsibility of clinicians from both health care services should decrease the challenges that these young adults face as the age-out of pediatric care. As discussed in the previous chapter, patients in the pediatric system receive stellar comprehensive care that is provided by a health care team. Despite the team approach, 20 participants (71%) indicated that there were no criteria for identifying patients who should be transitioned. Pediatric providers responded that they have provided care to patients as old as 25 years. There are no guidelines, written policies or tools to assess readiness for transition to adult care.

In the adult clinic, there is no dedicated social worker, allied health professional (nurse practitioner or physician assistant) or nurse to see patients with sickle cell disease. There is only one physician assigned to see patients with sickle cell disease, and the nurse and the social worker in the division cover all non-malignant hematological conditions.
Having a dedicated team specifically for patients with sickle cell disease would fill the service gap that now prevents patients from accessing care in the outpatient setting and encourages them to resort to emergency department care.

One barrier that is categorized as both a provider and system barrier is unique to the adult clinic. It is the requirement that all patients with sickle cell disease have a primary care provider. From a safety standpoint, since there are limited sickle cell disease clinical hours in the outpatient clinic, a primary care provider could be a valuable contact. However, findings from Chapter 4, which addressed patient barriers, suggest that primary care physicians are not comfortable managing patients when they are experiencing sickle cell related complaints. In addition, there is reluctance on the part of both hematologists and primary care physicians to prescribe narcotic pain medication for the patients when they are experiencing a painful crisis. The reluctance to take ownership of these patients is one of the major reasons that these young adults turn to emergency services, sometimes with much reluctance, for care with any acute illnesses.

Participants in all departments acknowledged that it is very difficult to transition patients to the adult system. Twenty-four participants (63%) acknowledged that there are difficulties regarding transfer of care from pediatrics to adult health care service. Apart from the pediatric emergency department, which has a hard age cut-off limit of 21 years, no other services in the children’s hospital have such a hard age limit when the pediatric provider will stop providing care to the patient. Overall, the varied pediatric specialists who provide care to patients with pediatric onset chronic illnesses have no specific age cut-off limitation.
Health care providers identified barriers in all categories across professions and departments. However, the findings from this study suggest that multiple system and provider barriers converge to explain why young adults continue to receive care in pediatrics. There are no written transition policies stipulating age limits or outlining the process of transition. In the pediatric system, there is no staff dedicated specifically to assist with transition; there are no comprehensive criteria for identifying those who should be transitioned, and there are no guidelines outlining the necessary steps to be taken to transition young adults from pediatric to adult health care services in a timely manner. Inadequate staff in the adult outpatient setting, delayed or inadequate transition preparation and a lack of resources were the most frequently identified barriers by health providers.

5.3.2.2. Access to Care

Timely access to preventive and acute care services for patients with sickle cell disease is important in preventing, maintaining and reducing the chance of disability and premature death. As stated above, access to age-appropriate care is delayed because patients remain in the pediatric system longer than they should. The system barriers discussed earlier, (lack of transition staff dedicated to individuals with sickle cell disease, long wait times for appointments in the adult system) all have an impact on access to care. The following responses from health care professionals who work in the emergency department indicate the burden of the problem regarding access to care for this population; “It takes FOREVER to get a new patient appointment,” “unable to find adult
“hematologist,” and “adult providers are not interested in caring for these patients because they take a lot of time.”

As discussed earlier, the most frequent reason for accessing acute care services is for pain. However, health care providers, both in the outpatient setting and the emergency department, harbor doubts about giving patients narcotic pain medication. The following are quotes from the survey of health care providers in the emergency department: “HCP [health care provider] prejudice that adult patient is drug seeking,” “adult providers are skeptical of painful crisis”, and adult provider is “less willing to give opiates.” Young adult patients are, therefore, at risk of experiencing delays in getting appropriate medical care because of possible stigmatization that they are “seeking opioids.” The current opioid crisis in the general population compounds the problem of opioid use for patients with SCD. Opioid addiction and overdose have led to increased barriers for patients with sickle cell disease. Sinha and colleagues (2019) reported that patients with sickle cell disease noticed that clinicians were more restrictive with opioid prescriptions and their clinicians’ focus on reducing their pain medication resulted in further stigmatization (Sinha et al., 2019). Several participants from the adult system also identified insurance as an access issue. However, this research did not include insurance as a variable.

5.3.2.3. Sickle Cell Disease

In terms of health care providers, there are established guidelines on the care of patients with sickle cell disease with acute complaints (Cantrill et al., 2012; Dowell et al., 2016). Judging from the responses provided by the participants that indicated that adult providers are reluctant to provide pain medications, there appears to be an underlying
stereotype that patients’ level of pain do not warrant opioid use or require the amount of opioid that is necessary to provide effective relief. This practice can result in patients receiving suboptimal care.

5.3.2.4. **Information Technology**

Information technology for health care providers include the availability and use of electronic medical records. The Health Information Technology for Economic and Clinical Health (HITECH) Act of 2019 was created to encourage health care providers and hospitals to use electronic health records (Menachemi & Collum, 2011). With this in mind, several questions (17 to 19) were framed to capture the use of information technology in managing the health of this population of patients. Health care providers across all departments indicated that there are multiple electronic medical record systems, and that all providers do not have access to all of them. For example, the emergency department clinician does not have access to the outpatient record despite being located on the same campus.

5.3.2.5. **Communication**

Effective communication is critically important, especially when providing health care services to this vulnerable population. There are multiple situations that can impact communication between patient and provider, and vice versa. Among them are intellectual deficits due to strokes in childhood. As discussed in chapter 2, SCD is the leading risk factor for strokes in black children (Baker et al., 2015). Immaturity in self-care choices and inability to articulate chief complaints are other examples of situations
that could impact communication. It was difficult to fully assess the degree to which health care providers experience barriers in communicating with patients. This is due to the fact that the questions about communication were closed-ended so the respondents could not elaborate. However, I deduced from the responses from some adult clinicians that there may be some impairment in communication as evidenced by remarks such as, “patients are not responsible for their own care,” and “patients should have written pain plan, similar to those who have asthma.” From the pediatric providers perspective, patients are not receiving education about transition.

5.4. Research Question 4

What are health care providers perceptions about how and when transition from pediatric to adult health services should begin?

The exploratory nature of the above questions identified some salient unmet health care needs for young adults with sickle cell disease. They are remaining in the pediatric setting long after they should have been incorporated into the adult system. This question aimed to elicit information about how to resolve the problem. A series of forced-response questions were used to gather input about the process of transition (questions 6, 7, 8 and 9), the staff required to overcome the challenge and those individuals who should work together to set priorities for development of a transition program.

Thirty-six participants indicated that transition should begin between age 15 to 21 years, one participant did not indicate the age at which transition should begin and another indicated that it should start between the ages 12-15 years. All participants
indicated that transfer to adult health care system should take place between the ages of 18 and 24 years. The general consensus across care settings was that planning should include both patient and health care provider representatives from pediatric and adult health care, parents, a social worker, a pharmacist, insurers and health care system administrators. Findings suggests that these health care providers recognize the problem; they have identified the necessary stakeholders to resolve the problem. Although it may be difficult to get this team together, the availability of web-based platforms like Zoom and Webex may make it easier to elicit input from this diverse group of individuals.

5.5. **Summary of Findings**

Health care provider data were collected using an electronic web-based electronic survey from 38 health care providers (RNs, NPs, and physicians) who work in the pediatric hematology clinic, the pediatric emergency department, the adult hematology clinic and the adult emergency department.

The answers to research question number 3 were categorized under the concepts outlined in the study. This categorical analysis identified three emergent themes related to individual barriers: the patients’ **unwillingness to transition to adult the health care system**, **fear and anxiety** about leaving the familiar pediatric setting and **trust of the pediatric provider**. **Intellectual deficit** was also identified as a barrier to timely transitioning into adult care. Pediatric providers did not identify any barriers that could be attributed to patients.
The health care provider barriers included pediatric providers’ reluctance to transition patients, pediatric providers’ inability to prepare young adults to manage their care, and inadequate of adult providers to provide care to this population.

System barriers were attributed to limited resources in the adult system, inadequate adult providers to provide hematological health care services, and no resources/staff for coordinating and providing transition services for young adults with such complex health care condition. A system barrier that is problematic and that can confer significant negative health outcomes is that of stereotyping patients as drug seeking.

In terms of access to care, there is excellent clinical care in the pediatric system but there were no criteria for identifying those individuals who should be transitioned and there are no policies on transition and no transition training. It is difficult to get appointments in the outpatient clinic partly as a result of limited resource allocation (less staff, both clinical and supportive), and partly as a result of limited hours of operation.

The findings regarding knowledge deficit were deduced from the responses of several emergency department clinicians. There are clinical treatment guidelines regarding timely evaluation of patients with SCD who presents to the emergency department. The guidelines include directives for the administration of pain medication with 30 minutes of evaluation (Ballas et al., 2012; Yawn et al., 2014). However, some health care providers identified resistance to prescribing pain medication as a barrier to care. Patients perceive this resistance to prescribing narcotic pain medicines as stigmatization, and implied that this bias experienced quite often in the emergency
department. Emergency department clinicians are accustomed to following guidelines for treatment of various ailments such as management of patients presenting with chest pain, infection, and strokes. Therefore, it should not be burdensome for health care providers to follow clinical practice guidelines for treatment of patients with SCD that were released by the American Society of Hematology (Brandow et al., 2020). These guidelines, if implemented, will alleviate the issues of having patients with SCD wait for prolonged periods of time to be evaluated and to receive medication for pain.

Health care providers indicated that effective communication is hampered by the use of **multiple electronic medical record platforms that are not accessible to all clinicians**. Each department uses a different electronic medical record platform. Health care providers do not have access to medical records across platforms because each platform requires personal, department protected password to enter patients’ medical records. Effective communication is the cornerstone of safe, effective care, whether it is between patient and provider or provider and provider.

A number of interrelated themes emerged from this study. They highlight the reasons for the delay in transitioning young adults with sickle cell disease from the pediatric to the adult system. Findings from the forced-response questions suggested that transition should begin around the age of 15 years and transfer of care should occur around the age 21 years. Respondents (patients and health care providers) expressed some degree of flexibility regarding the upper age limit when patients should be fully transitioned from the pediatric system. Multiple providers suggested that by age 24 years, patients should be receiving all their care in the adult system.
The resolution of the problem of young adults remaining in pediatric care longer than they should include the involvement of multidisciplinary stakeholders such as patients/family, administrators, clinicians from both pediatric and adult systems, pharmacist and social work. It is within this context that a proposal for a transition model is made to bridge the gap in care. This model is described in Chapter 7.
CHAPTER SIX

CONCLUSION: REVIEW OF PROBLEMS AND RESEARCH QUESTIONS

6.1. Review

Sickle cell disease (SCD) is the most common genetic disorder in the United States. Although it can be found in other ethnic groups, it primarily affects individuals of African descent (Carden & Little, 2019; Neumayr et al., 2019). Sickle cell disease is characterized by anemia and a constellation of acute and chronic complications that result in increased morbidity and mortality. However, recent advances in medical technology, diagnosis and medical management have resulted in extending life expectancy from the early teens into the fourth and fifth decades. More than 93% of children now survive to adulthood (Quinn et al., 2010). As a result, although SCD was once considered a health condition of childhood, it now poses challenges for young adults who have survived the illness and who must now transition from the pediatric to the adult health care system.

This study explored the patient, health care provider and health care system factors that impact transition of young adults with sickle cell disease. Participants identified barriers that explain why young adults with SCD are still receiving their health care services in pediatrics. To my knowledge, this is the first study to examine the issue holistically by including patients and providers in a single tertiary care facility that has the infrastructure to provide sickle cell disease specialty care to both pediatric and adult patients. Creating comprehensive transition pathways for young adults with SCD ensures that they continue to receive optimal health care services. This is particularly important because young adults are at highest risk for early mortality (Quinn et al., 2010; Hamideh & Alvarez, 2013). A single-center cohort study conducted by Quinn and colleagues
(2010), found that majority of deaths that occurred in individuals over the age of 18 years occurred after transfer to an adult provider. According to a statistical brief from the Healthcare Cost and Utilization Project sponsored by the Agency for Healthcare Research and Quality, and the results of a retrospective cohort of SCD-related emergency department visits and readmissions from eight geographically dispersed states, sickle cell disease had the highest 30-day readmission rates for individuals aged 18 to 30 years old (Brousseau et al., 2010b). The hospital readmission rate was up to 48.3% more than other individuals in the same age group. That is more than that of any other medical condition (Brousseau et al., 2010b; Elixhauser & Steiner, 2013). These findings highlight the critical need for patients with SCD to have effective transition pathway from pediatric to adult healthcare system.

As described in detail in Chapter 1, and briefly reiterated here, health care transition was defined more than 20 years ago as “the purposeful planned movement of adolescents and young adults with chronic medical and physical conditions from child-centered to adult-centered health care services” (Blum et al., 1993). In 2002, the American Academy of Pediatrics (AAP), the American Academy of Family Physicians (AAFP), and the American College of Physicians – American Society of Internal Medicine developed a consensus statement on healthcare transition for adolescents and young adults with special health care needs:

“The goal of this policy statement was to ensure that by the year 2010 all physicians who provide primary care or subspecialty care to young people with special health care needs would understand the rationale for transition from child-oriented to adult-centered care; have the knowledge and skill to facilitate that process; and know if, how and when transfer of care is indicated” (AAP, 2002b, pp. 1304).
However, decades after the target date for achieving that goal, the health care system continues to struggle with how to create effective pathways to transition from pediatric to adult health care services for young people with chronic illnesses like sickle cell disease. The paucity of evidence-based guidelines concerning the best practices for successful transition from pediatric to the adult health care system for young adults with sickle cell disease was the catalyst for undertaking this mixed-method research study.

The qualitative data gathered through individual interviews and memos provided detailed information about the problem by emphasizing the voices of patient participants. Twenty-four individuals signed consent forms and 16 (67%) individuals, ranging in age from 18 to 30 years, were interviewed. The quantitative portion gathered descriptive information from 38 health care providers who worked in the pediatric hematology clinic, the pediatric emergency department, the adult hematology clinic and the adult emergency department. They provided information on the current state of medical care for these patients, their perceptions of barriers and/or facilitators of transition and recommendations for seamless transition for this population of patients by completing web-based surveys.

In this study, concepts of the Patient-Centered Medical Home Model (PCMH) and elements from the National Quality Forum Framework (NQF) for Measuring Care Coordination were used as a guide to frame and develop the research questions. Both the PCMH and the NQF were discussed in Chapter 2. To briefly reiterate, the PCMH is designed around the needs of the patient with chronic illnesses. It is defined as the “provision of comprehensive primary care services that facilitates communication and shared decision-making between the patient, his/her primary care providers, other
providers, and the patient’s family” (American Hospital Association, 2010). The complexity of SCD management and the need for multi-disciplinary services made using elements of the PCMH framework essential in framing the questions that provided the answers to the larger research question.

The National Quality Forum Framework for Measuring Care Coordination (NQF) describes the basic elements needed for optimal transition of care in terms of structural quality; the common processes that should occur in any setting of care and that can be applied to all patients experiencing transition. The NQF describes health care coordination as a process “that ensures that patients’ needs and preferences for health services are met.” It also advocates that there should be effective communication between the patient and family, health care provider, proper health maintenance (surveillance and prevention) and facilitation of information sharing across health care disciplines and sites (NQF, 2006). To this end, elements of both frameworks were used to guide the design of the interview guide for patients and the survey for providers in both the pediatric and adult healthcare systems. This comprehensive approach provided an opportunity to understand the issues that are critical to timely, successful transition for young adults with sickle cell disease.

The following research questions and findings provided insight into the extent of the problem of why young adults with SCD continues to receive health care services in pediatrics. It also provided guidance for the resolution of the problem.

1. What are the individual, provider and system factors that patients perceive as facilitating and/or impeding transition from pediatric to adult health care system for young adults with SCD?
2. Are young adults with SCD prepared to access and navigate the adult health care system?

3. What are health care providers’ perception of patient, system and provider factors that impact transition from pediatrics to adult health care system?

4. How and when should transition from pediatric to adult health care services begin?

The methodology for examining each of these questions is detailed in the methodology section in Chapter 3. Briefly, to better understand the state of transition and the factors that impact transition from pediatric to the adult health care setting individual interviews with patient participants were used to answer Questions 1 and 2. Electronic web-based survey was used to answer Question 3. Responses from the open-ended questions on the electronic web-based survey and from the patient participants’ interviews provided the answers to the last research question. Although data collection was done simultaneously, data collection from patient participants occurred over a longer period of time.

Five concepts generated from the research questions and a review of the transition and sickle cell literature formed the basis for the organization of the analyses. The key concepts are transition, access to adult health care services, sickle cell disease, information technology and communication. A thematic framework that was developed around elements of the Patient Centered Medical Home model (Agency for Healthcare Research and Quality) and the National Quality Forum (Antonelli et al., 2009; National Quality Forum, c2021) framework for measuring care formed the template for organizing codes and guiding data classification.
6.2. Discussion

This study found that multiple sources (patient, provider and system) are responsible for causing young adults with SCD to remain in the pediatric setting longer than they should. As discussed earlier, transition is a process that occurs over time. Multiple studies have focused on barriers to care and have reported similar findings (Stollon et al., 2015; Sobota et al., 2015; Gray et al., 2017). However, this study expands on the scope of previous studies. It presents a more comprehensive examination of why young adults with sickle cell disease continue to receive health care services in pediatrics when they should be in the adult system. It reports the findings from the perspective of patients with SCD in both pediatric and adult health care systems, as well as health care providers from adult and pediatric sickle cell clinics and emergency departments in the same tertiary health care facility.

The key findings of the study are summarized and reported according to the research questions. The factors that can be classified as facilitators of transition were sparse; only one participant was able to articulate a facilitating factor as described in Chapter 4. In response to research Question 1, the patient participants identified several factors that explained why it was difficult to leave the pediatric health care system when they reached the age of legal majority. The patient participants reported the following as individual barriers: lack of knowledge about transition, sickle cell disease and the complications of sickle cell disease; trust of their pediatric health care providers; comfort with the care that is provided in the pediatric setting (acute and routine); fear of leaving pediatrics; assuming full responsibility for their own care; and perceived isolation in the adult setting.
The participants who are receiving their health care services exclusively in the pediatric system did not articulate barriers that could be attributed to health care providers. However, those patient participants who have already made contact in the adult system, either by using the emergency department because they are 21 years and older or have been to the adult clinic reported that they received minimal or no preparation for transition to the adult system. All participants who have experienced care in the adult system articulated that there are major differences in the way the care is delivered. For example, when they access the emergency department for an acute illness, they are immediately evaluated in the pediatric emergency department. However, in the adult emergency department, they oftentimes experience long wait times to be evaluated and even longer wait times to receive medication for pain.

Some of the barriers that were identified by participants aged 21 years and older can be classified both as provider and system barriers. There are inadequate numbers of health care providers and supportive staff in the adult health care system and insufficient appointment slots to accommodate patients in the adult clinic. There is also inadequate information about SCD and transition and an absence of an organized transition program.

Evidence from the literature shows that pediatric health care clinicians provide excellent health care services using the family-centered approach (Kuhlthau et al., 2011). Additionally, the literature shows that pediatric health care providers’ adherence to clinical guidelines and evidence-based practice plays a major role in the survival of children (Cabana et al., 2019). Recent advances in science and medical technology are now positioning individuals with sickle cell disease to benefit from such inventions as
gene therapy and medications that ameliorate symptoms and improve quality of life for these patients (Salinas Cisneros & Thein, 2020; Ballas & Darbari, 2020). However, in order to continue to narrow the morbidity and mortality gap, it is important to integrate transition planning in routine care so that pediatric patients have adequate time to learn about SCD and its complications, develop the skills that are necessary to navigate the adult health care system and participate in the management of their health. This comprehensive approach undoubtedly would result in improved quality of life and decreased morbidity and mortality.

Findings related to the second research question suggest that knowledge and preparation confers some degree of empowerment that positions an individual to be more involved in his/her care. Most of the participants had limited knowledge about sickle cell disease and its complications. Fifty-six percent of the study population were able to name three complications of sickle cell disease and three of them were not able to identify any complications. Education level did not make much difference in knowledge about sickle cell disease. There were college students who were not able to articulate as much knowledge about sickle cell as non-college educated participants. However, in this study, the participants who had less knowledge about sickle cell were less consistent with getting routine preventative care.

Participants who were receiving care (emergency and routine) in the adult system reported that they had minimal or no preparation about what to expect before they ended up in the adult system. One possible explanation for these findings is the fact that there are no established curricula, protocols or guidelines to aid or guide the process of transition. Study participants see transition as a process; however, they reported that some
providers only mentioned that “someday” they would go into the adult system, and that was where the conversation ended. As a result, they lacked the skills to competently navigate the adult system.

Pediatric providers have the trust of their patients because of long-established relationships, some from infancy. Additionally, the structure of care in the pediatric setting also has the necessary staff (counsellor, child life specialists, nurse practitioner, dietician and social worker) to integrate transition planning in care. However, incorporating successful transition requires adopting a clinical model that incorporates a comprehensive, holistic approach that addresses the physical, psychological and social needs of the patient. Such an approach has the potential to deliver significant health benefits, while at the same time providing cost savings in terms of personal earnings and decreased use of emergency services. Those patients who are employed may have more productive working days and overall better quality of life.

As outlined in the Chapter 3, answers to research Question 3 were elicited by web-based survey. They identified patients’ level of maturity, parental control, parents and patients’ refusal to leave pediatrics and intellectual deficits as individual barriers to transition. Of note, pediatric providers attributed no patient level barriers that impacted transition. Nurses identified similar individual barriers (trust of pediatric providers, fear of the adult system) to patients’ not leaving pediatrics. However, the lack of staff in the adult outpatient setting, delayed or inadequate transition preparation, lack of resources for transition services and support in the adult system, and multiple computer systems that are not accessible to all providers were the most frequently identified barriers.
In response to Question 4, findings from both patients and providers were similar. Thirty-six participants indicated that transition should begin between the ages of 15 to 21 years, one participant did not indicate the age at which transition should begin and another indicated that it should start between the ages 12-15 years. All participants indicated that transfer to adult health care system should take place between ages 18 and 24 years. The general consensus across care settings is that planning should include both patient and health care provider representatives from pediatric and adult health care, parents, a social worker, a pharmacist, health insurers and health care system administrators. Findings suggest that these health care providers and patients recognize the problem and have proposed similar solutions. They have also identified similar stakeholders who are necessary to resolve the problem.

6.3. Other Findings

Other critical findings emerged from this study that contribute to knowledge about the current state of care for patients with SCD. The National Heart, Lung, and Blood Institute (NHLBI) has published guidelines for the management of pain in SCD crisis. Patients report that they sometimes delay accessing health care services because they feel that they are “judged as drug seekers.” Patient participants perceived that clinicians in the emergency department and the outpatient settings (primary care and hematologists) are hesitant to prescribe narcotic pain medications when they are experiencing pain crises. The following responses from emergency department physicians, “adult providers are skeptical of pain” and “health care providers prejudice that the patient is drug seeking,” seem to corroborate the patients’ perceptions that adult
clinicians “judge them as drug seekers.” The perception that health care providers do not believe in the degree of pain that patients are experiencing is partly responsible for negative attitudes towards adult health care providers, and the way patients sometimes delay accessing health care services until they in severe distress. Patient participants perceive that adult health care providers are reluctant to provide adequate medications for pain. Findings from this study suggests that this underlying bias can result in patients receiving suboptimal care.

6.4. **Limitations of Research**

One of the primary limitations to this study is the small sample size of 16 patients and 38 health care providers. The findings are not generalizable because the study was conducted in a single institution. Repeating this study in multiple tertiary care facilities where there is co-location of pediatric and adult hematologists would clarify the generalizability of the results. The study was conducted only among individuals who are English-speaking and among those who were receiving medical care. Including those individuals who were not English speaking and who were not receiving routine care would add more strength to the findings because they may experience additional barriers that are not experienced by those who participated in the study.

Another limitation of this study is recall bias, because the majority of the patients who were in adult care reported that they were not prepared to enter the adult setting. Although the questions for the interview were developed from a review of the literature, the omission of important factors like having health insurance and reliable transportation may have significant negative impact on access to health care services.
6.5. **Programmatic and Policy Implications**

In order to effectuate improvement in care for young adults with SCD, programmatic and policy changes are necessary. This study suggests that the following programmatic and policy changes could help to improve transition for young adults with SCD from pediatric to the adult health care system. They could also improve access to SCD specialty care and the quality of care that patients receive in the adult health care setting.

The programmatic and policy recommendations emerged as a result of integration of the findings from both study populations. However, for these programmatic and policy changes to be effective, it is detrimental to get the support of operational and administrative principals. Hospital and clinic administration, health insurance representatives and private/philanthropic organization funding sources are important allies to partner with patient representatives, pediatric and adult hematologists, emergency medicine providers, pharmacists, social workers and community partners to remove the barriers that prevent young adults from crossing the pediatric to adult health care services chasm. Once these barriers are removed, the fear that young adults now experience, will be significantly diminished or eliminated.

The cut-off age for pediatric patients to be seen in the pediatric emergency department is 21 years. Including transition planning as part of anticipatory guidance in pediatric care provides an excellent opportunity to provide sickle cell disease education and guidance for this population of patients. Table 6 summarizes some of the immediate programmatic changes that could be implanted and proposes long-term policy changes.
that can positively impact successful transition from pediatric to adult health care services.

Table 6: Programmatic & Policy Recommendations for Improving the Care of Adults with SCD

<table>
<thead>
<tr>
<th>Programmatic Changes</th>
<th>Policy Changes</th>
</tr>
</thead>
<tbody>
<tr>
<td>1) Develop a transition blueprint indicating goals, when transition should begin and a pathway to follow</td>
<td>1. Develop transition policies that give clear guidelines (across specialties) regarding the process, timing and evaluative processes. Policies should be easily accessible to all health care providers.</td>
</tr>
<tr>
<td>2) Develop education curriculum and standards to measure acquisition of knowledge</td>
<td>2. Include sickle cell management as part of the curriculum in the annual grand rounds schedule for emergency department faculty</td>
</tr>
<tr>
<td>3) Set individual goals with patient and family.</td>
<td>3. Conduct quality improvement project to address “time to administration of pain medication” for those individuals who present with a pain crisis. Use the results to develop a pain management protocol so that patients do not have to wait for long periods of time to be evaluated by a clinician, and then wait even longer for administration of pain medication</td>
</tr>
<tr>
<td>4) Conduct workshops and provide opportunities for patients to roleplay</td>
<td>4. Use of a single computer system at RWJBH so clinicians will have access to medical records</td>
</tr>
<tr>
<td>5) Incorporate symbolic graduation prior to transfer to the adult health care setting.</td>
<td></td>
</tr>
<tr>
<td>6) Provide opportunities for group projects.</td>
<td></td>
</tr>
<tr>
<td>7) Develop individual health passports that have pain management plan.</td>
<td></td>
</tr>
</tbody>
</table>
CHAPTER SEVEN

THE BRIDGE MODEL

7.1. BRIDGE Model of Transition

The American Society for Adolescent Medicine describes transition as an active medical process, “the purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from a child-centered to adult-centered healthcare systems.” (Blum et al., 1993). Getting from one place to another oftentimes involves crossing a bridge and it is within this context that the BRIDGE Model of Transition is introduced.

![Figure 10: The BRIDGE Model of Transition](image)

Pediatric providers form the pillar at one end and adult providers form the pillar at the other end of the BRIDGE Model of Transition, while the nurse case manager (NCM) serves as the guide who coordinates appropriate health care services and liaises with other members of the health care team. An allied health professional (such as a family nurse practitioner or a physician assistant) and social worker are the girders of the BRIDGE Model of Transition.

The model proposes a transition plan that utilizes a comprehensive multidisciplinary approach. Adolescents and young adults receive individualized
education about sickle cell disease, potential complications of sickle cell, medications, and symptom management starting at around the time they enter middle school. The plan follows a set curriculum that can be customized to account for the cognitive function, literacy and cultural mores of the patient and family. There are existing transition tools which can be accessed through GotTransition via the organization’s website at: www.gottransition.org, and they can be adopted to meet the unique needs of the patient population.

Medical care in pediatrics utilizes a family-centered approach, while an individualized approach is practiced in the adult setting. It is within these individualized health care settings that young adults experience isolation and confront barriers and challenges of navigating the adult system of care. Social workers are uniquely positioned to assist these individuals to adjust and develop the necessary skills to navigate the challenges of a health care landscape that is oftentimes difficult to travel, especially for individuals with pediatric onset chronic illnesses. Social workers have the training to address the complex challenges that these individuals face. They also have access to a wealth of community resources, linkages and partnerships to assist young adults with SCD to achieve the best medical and social outcomes. Other relevant problems that social workers are qualified to address include housing, employment, disability, substance abuse and insurance issues that young adults with sickle cell disease oftentimes confront (Saxe Zerden et al., 2019).

A nurse case manager is a Registered Nurse (RN) whose primary role is to coordinate health care services, supervise and maintain responsibility for clinical care and facilitate communication between the clinical providers and patients (Reimanis et al.,
In the *BRIDGE model*, the NCM assists with patient education, medication adherence and ensures that patients receive the resources and education that are required to competently function in the adult health care system. The nurse case manager will also serve as the liaison between hematologists, primary care providers and other subspecialists.

In the proposed model, an allied health professional (family nurse practitioner or physician assistant) will work collaboratively with the pediatric and the adult hematologists. This professional has the training to assess patient needs, order and interpret diagnostic tests, diagnose illnesses, formulate and prescribe treatment plans. The allied health professional also has the training and licensure to work collaboratively with pediatric and adult hematologists.

As the patients gradually prepare to enter the adult health care system, the allied health professional who has training and the licensure to provide clinical health care services to individuals in both pediatric and adult populations, the social worker and nurse case manager who works with them in the pediatric setting cross over with them to the adult setting. The social worker and the allied health professional are girders of the transition *BRIDGE Model of Transition*. They, along with the nurse case manager who serves as the guide, escort transitioning patients from pediatric to adult health care services.

Findings from the study suggest that consistency and trust are important constructs for young adults who are transitioning from pediatric to adult health care system. Having existing relationships with trusted health care providers as they make the trajectory across the *BRIDGE* into the adult setting confers a degree of support that is
essential to prevent disconnection from primary preventive care. It also empowers transitioning patients to receive the care that they need to improve their quality of life, reduce emergency department visits and ultimately decrease morbidity and mortality.

7.2. Acknowledgement of Research Study Participants

Thank you to those patients with sickle cell disease whose unbound resilience to tackle the challenges of life inspire me despite the way that you sometimes endure the burden of significant pain. Your participation is deeply appreciated. Your voices have shed so much light on the struggles that so many people with sickle cell disease experience. Thanks for your openness and honesty in helping us understand this problem. Moreover, thanks also for your recommendations regarding how to resolve these problems.

Thanks to all the clinicians who took the time out of their busy schedules to share their opinions and make recommendations about how to improve medical care for patients with sickle cell disease. Your recommendations helped to inform the development a quality improvement project aimed at decreasing the time for a patient to be evaluated and given pain medication based on the time they were triaged in the emergency department. Your recommendations also helped to inform a proposal for resolving the issue of transition for young adults with sickle cell disease.
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APPENDICES

APPENDIX I: Sickle Cell Disease Research Study

SICKLE CELL DISEASE RESEARCH STUDY

• Looking for individuals with sickle cell disease to participate in a research study to find out about their experiences with transitioning and transferring from pediatric care to adult care.

• To participate in the study, you must have sickle cell disease and be between 18 and 30 years of age.

• You will be interviewed either in-person or by telephone to try to understand your experiences with preparation, transition and transfer of care from the pediatric to the adult health care system. The interview will take about 30 minutes and you have the option to agree to or decline audiotaping.

• All information will be confidential. Your name will never be recorded or used.

• After completing the interview, you will receive a $25.00 gift card as an incentive for participating in the study.

Principal Investigator: Paulette Forbes, PhD candidate
Rutgers University/New Jersey Institute of Technology
Joint PhD in Urban Systems Program
65 Bergen Street, Room 1127, Newark, New Jersey 07107
Contact: forbesp2@cnj.rutgers.edu
Telephone: 732-853-4245

Rutgers, The State University of New Jersey
Protocol Version - 1.0 March 25, 2016
APPENDIX II: Consent to Take Part in a Research Study

CONSENT TO TAKE PART IN A RESEARCH STUDY
In-Depth Interviews

TITLE OF STUDY: “Why are Young Adults with Sickle Cell Disease Still in Pediatric Care?”

Principal Investigator: Paulette E. Forbes, PhD candidate

This consent form is part of an informed consent process for a research study. It will provide information that will help you decide if you wish to participate in this research study. It will help you understand what the study is about and what will happen during the course of the study.

If you have questions at any time during the consent process, feel free to ask them. You should expect to be given answers that you completely understand. After all of your questions have been answered, if you still wish to take part in the study, you will be asked to sign this informed consent form. You are not giving up any of your legal rights by volunteering for this research study or by signing this consent form. You will be given a copy of the signed consent form to keep.

Who is conducting this research study?
Paulette Forbes is the Principal Investigator (PI) of this research study. The PI has the overall responsibility for the conduct of the study. However, there are often other individuals who are part of the research team.

Who might benefit financially from this research?
No one will gain any financial benefit from this research study.

Why is this study being done?
To understand the factors that impact health care transition from pediatric to adult health care services for young adults with sickle cell disease.

Why have you been asked to take part in this study?
You are being asked to participate in the study because you have sickle cell disease and you are between the age of 18 and 30 years.

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Fax: (973) 972-3225
Who may take part in this study? And who may not?
Individuals who have sickle cell disease and speak and understand English. Individuals with cognitive and mental disorders that may interfere with understanding the informed consent process or their recollection of past events will not be asked to participate.

How long will the study take and how many subjects will participate?
The study will last up to six months. Up to 45 participants will be recruited to participate in interviews. Each interview is expected to last between 30 and 45 minutes.

What will you be asked to do if you take part in this research study?
You will be asked to share your experience with getting sickle cell disease care as you reach adulthood and what you think are the factors that impact the movement from pediatric to adult health care services. The interviews will be recorded. If you choose not be audiotaped, notes will be taken during the interview. All interviews will be done privately. You have the option to conduct the interview by telephone if in-person is not convenient.

What are the risks and/or discomforts you might experience if you take part in this study?
The principal risk would be a breach of confidentiality. Because of this risk, your comments/discussions will be kept confidential and will not be reported with any information that could identify who you are. All audiotapes and notes will be locked in a secure place and will be destroyed at the completion of the study.

Are there any benefits for you if you choose to take part in this research study?
You may not receive any direct personal benefit from taking part in this study. However, the results have the potential to provide the blueprint for the creation of an effective transition model. This model of care will ensure that adolescents and young adults with SCD move seamlessly from pediatric to adult services utilizing a coordinated approach.

Will there be any cost to you to take part in this study?
There will be no cost to you for participating in the study.

Will you be paid to take part in this study?
You will receive a $25.00 gift card for taking part in the study after completing the interview. If the interview is conducted on the telephone, you may collect the gift card in person, or it can be mailed to you.

How will information about you be kept private or confidential?
To ensure that confidentiality is maintained at all times, participant data will not include name, date of birth, telephone number or address. Pseudonyms will be assigned to all participants. Signed consents will be kept in a separate folder in a locked cabinet in the PI’s lockable office.
What will happen if you do not wish to take part in the study or if you later decide not to stay in the study?
Participation in this study is voluntary. You may choose not to participate, or you may change your mind at any time and withdraw from the study.

Who can you call if you have any questions?
If you have any questions about taking part in this study, you may contact:
Paulette Forbes, PhD candidate
Joint Urban Systems Program, Urban Health
Contact Number 732-853-4245

If you have any questions about your rights as a research subject, you can call:
The IRB Director: (973)-972-3608 Newark or
Human Subject Protection Program: 973-972-1149 - Newark

What are your rights if you decide to take part in this research study?
You have the right to ask questions about any part of the study at any time. You should not sign this form unless you have had a chance to ask questions and have been given answers to all of your questions.

AGREEMENT TO PARTICIPATE
I have read this entire form, and it has been read to me, and I believe that I understand what has been discussed. All of my questions have been answered.

Subject Name: _______________________________

Subject Signature: __________________________ Date: ______________

Please circle and initial your preference for recording the information from your interview
Audiotape  No audiotape

Signature of Principal Investigator:

I have explained the full contents of the study to the above named participant to the best of my ability. All the participant’s questions have been answered accurately.

Principal Investigator: Paulette Forbes, PhD candidate

Signature: ____________________________ Date: ______________
CONSENT FOR THE USE OF AUDIOTAPE RECORDER

As part of this research project, audio recording will be made of my interviews. Only members of the research team will listen to the recordings. The recording will be transcribed word-for-word and anything that can identify who you are will be deleted.

I understand that I can refuse to be audiotaped and still participate in the study.

I ________________ authorize audiotaping of interview subject to the following conditions:

1. I understand that I will not be identified in any transcription or publication using information from the interviews.
2. I understand that the research team and the IRB have approved the use of audiotape.
3. I understand that all recordings will be destroyed after the interviews have been transcribed.
4. I understand that the tape-recorded interviews will not be used for commercial or public media purposes.

SIGNATURE OF SUBJECT

Signature: ___________________________ Date: ________

Rutgers, The State University of New Jersey

Protocol Version – 1.0 March 25, 2016
APPENDIX III: Interview Guide – Patients Participants

Date: 
Pseudonym:  
Age: 
Educational level:  
Gender:  
Employment status:  
Place of care:  
Hemoglobin phenotype:  
Insurance status:  

**Introduction**

- Thank participant for agreeing to participate in the study  
- Review the purpose of the study  
- Describe the structure of the interview (audio-taping and note taking)  
- Provide information that no names will be used, how confidentiality will be maintained  
- Review information that participation is voluntary  

**Accessibility to Care**

- Do you have a regular hematologist? If no, how do you get sickle cell care?  
- How long have you been going to this provider?  
- In the last 3 months have you seen your provider for routine care? Not sick or follow-up care  
- How easy/difficult is it for you to get an appointment for the time that you wanted? Rate it on a scale of 1. Easy, 2. Not so easy, 3. Difficult – Explain  
- When you go for your appointment, how long is your wait time?  
  - What can be improved?
1. Share what you know about sickle cell
2. Can you tell me how you felt the first time you realized that sickle cell disease was going to impact your life for a long time
   a. Describe the experience you mentioned
   b. What was it like for you?
   c. Do you know what kind of sickle cell you have?
   d. What are some of the complications that an individual can get as a result of having SCD?
   e. Do you have any complications as a result of sickle cell disease?
   f. Do you know your medications (if any) including doses and allergies

Continuity of care

1. If you were to be admitted to a hospital that no one knows you, how confident are you to tell the healthcare provider how to care for you?
2. What would make you worry that the approach to your care is not standard of care for someone with SCD?
   a. Follow-up: Please give an example if you have ever had such an experience
3. Do you know what medicine works best for you when you are in crisis?
4. How confident are you to inform your health care provider what medicines work best for you when you have pain crisis?
5. Describe your experience the last time you were in the hospital
   a. What was the experience like for you?
   b. Have you been to the emergency room alone?
   c. If so, describe your experience
   d. Do you feel that you had a say in your care? Explain
6. Do you see your Dr. or NP alone?
7. Do you recall how old you were when you started to see your health care provider alone?
8. Who makes your follow-up appointments?
9. Do you call and cancel appointments if you cannot make it?

### Questions about transition & the healthcare system

1. Tell me, what do you know about transitioning from pediatric to adult healthcare service?
2. Did/Have your health care provider talk (ed) to you about going to the adult clinic? Describe
3. Were there problems? Describe
4. Do you think anything can be done within the healthcare system that would help you make the change easier?
5. Do/Did you feel prepared to leave the pediatric clinic?
   a. Yes, explain why
   b. No, explain why
6. Describe what you would like to see with regard to preparation for the adult clinic
7. When was the last time you were admitted to the hospital?
   a. Were you in the children’s hospital ER or the adult hospital ER?
   b. What was your experience?
   c. What if you were sent to the adult ER because you are too old for the Pediatric ER? Would that engender fear? Please describe how you would feel?
   d. What was your experience like the first time you went to the adult ER?
8. How do you feel about transition? What does it mean to you?
   a. When do you think it should occur? Explain
   b. Did/Does anyone in pediatric clinic discuss it with you? How old were you when it was first discussed?
   c. Do/Did you feel prepared? Why? Why not?
9. Describe what the ideal transition program would look like for you.
Information Technology

1. Do you have a smart phone?
2. Do you use it to help manage your care? Medication reminders, medication list, allergies, your last appointment, what medicines work best for you
3. Do you have access to your test results?

Quality and Safety

1. In terms of SCD, what is most important to you with regards to the future? Describe
2. What advice would you give to others who are in your position to make the changes?
3. Why do you feel that way?

Throughout the interview, various probes will be used for completeness. Examples include:

- Can you tell me more about that experience?
- Anything else?
- Can you be more specific?

Rutgers, The State University of New Jersey

Protocol Version – 1.0 March 25, 2016
APPENDIX IV: Email to Healthcare Providers

Email to healthcare providers: MDs, NPs, PAs and RNs in the Adult Hematology and Pediatric Hematology/Oncology clinics and the Pediatric and Adult Emergency Departments

I am a graduate student in the joint PhD program at New Jersey Institute of Technology/Rutgers University. My area of concentration is urban health. I am conducting a study to try to understand why young adults with sickle cell disease are still being treated in pediatrics. I would also like to find out how did young adults with SCD who are being treated in the adult system move over from the pediatric system. This information will assist in identifying the factors that impact transition of care and will assist in developing an effective transitional care model so that young adults with childhood onset chronic disease like sickle cell anemia will not experience any lapses in care, will not seek episodic care in the emergency department, and at the same time, they will receive age-appropriate care.

Thank you for agreeing to help me understand this very important issue by completing this survey.
APPENDIX V: Survey for Healthcare Providers

Survey for Healthcare Providers

1. Which of the following best describes your position?
   - MD-ED
   - MD-adult hematology
   - NP-ED
   - PA-ED
   - MD-pediatric hematology/oncology
   - NP-pediatric hematology/oncology
   - RN-adult hematology
   - RN-pediatric ED
   - RN-adult-ED

   Male  female

2. Is there a written policy on transition and transfer of care in your department?
   - Yes
   - No

3. Is there an age limit as to when patients are seen or not seen in your department?
   - Yes
   - No

4. Is there someone who explains the process of transition and transfer from pediatrics to adult healthcare services?
   - Yes
   - No

5. At what age do you believe the transition process should begin?
   - 12-15
   - 15-17
   - 18-21
   - Other

6. Are there criteria for identifying young adults who are in the process of transitioning?
   - Yes
   - No
   - NA

7. Do you have a list of adult healthcare providers to refer youths in case they do not have a provider?
   - Yes
   - No

8. At what age should transfer occur?
   - 18-21
   - 22-24
   - Other
9. Can you estimate the age of the oldest person with sickle cell disease that you have provided care for in the last year?

10. How difficult is it to transfer health care from pediatric services to adult services?
   1. Very difficult
   2. Moderately difficult
   3. Somewhat difficult
   4. No problem to transfer care
   5. N/A

11. Do you send a written summary to the accepting service?
    Yes  No  Sometimes  N/A

12. Is there anyone in your practice that communicates with adult providers prior to and after transfer of care?
    a. Yes – Who (use title, not name)
    b. No -

13. List four things that you perceive as barriers to transitioning young adults from pediatric to adult health care services

14. Please give four suggestions as who should sit at the table to discuss planning and implementation of transition program

15. What are some changes that you would like to see occur from the administrative level that would be supportive of a transition program? List as many as you wish.

16. Do you have access to electronic medical record across specialties? If no, please list suggested changes that you would like to see. If yes, please list what you are satisfied with and what can be improved.

Thank you for participating in this survey. Your feedback is important
### APPENDIX VI: Summary of Themes from Survey

<table>
<thead>
<tr>
<th>Research Question 3</th>
<th>Concepts</th>
<th>Codes</th>
<th>Themes</th>
</tr>
</thead>
<tbody>
<tr>
<td>What are health care providers’ perceptions of patient, system and providers’ factors that impact transition from pediatric to adult health care system for young adults with sickle cell disease?</td>
<td>Transition</td>
<td>Barriers to transition</td>
<td>Unwilling to leave pediatrics Trust/attachment Anxiety/fear Maturity</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Individual</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Provider</td>
<td>Lack of adult providers Not teaching patients to be responsible Refusal to transition pediatric patients</td>
</tr>
<tr>
<td></td>
<td></td>
<td>System</td>
<td>Lack of adult providers Reluctance to prescribe pain med Difficulty transferring care to adult heme Inadequate resources</td>
</tr>
<tr>
<td></td>
<td>Access to care</td>
<td>Pediatric care</td>
<td>No criteria for identifying who and when to transition</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Policy on transition Staff for transition</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Adult care Getting appointment Referrals/Resources</td>
<td>Difficulty transferring care to adult heme Cannot get adult outpatient appointments</td>
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<td></td>
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<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Sickle cell disease</td>
<td>HCP knowledge</td>
<td>Knowledge deficit</td>
</tr>
<tr>
<td></td>
<td>Information technology</td>
<td>Access to electronic medical records</td>
<td>No access to medical records because of multiple electronic medical record systems</td>
</tr>
<tr>
<td>Communication</td>
<td>Provider-to-provider</td>
<td>Patient-to provider</td>
<td>Provider-to-patient</td>
</tr>
</tbody>
</table>
### APPENDIX VII: Summary of Barriers to Transition

<table>
<thead>
<tr>
<th>Department/Profession</th>
<th>Findings</th>
<th>Barriers</th>
<th>Summary</th>
</tr>
</thead>
<tbody>
<tr>
<td>Emergency Dept</td>
<td></td>
<td></td>
<td>Both participants indicated fear of leaving pediatric care and lack of resources to support a transition program as barriers</td>
</tr>
<tr>
<td>RNs Adult (N=2)</td>
<td>1. Fear of leaving pediatric provider</td>
<td>1. I</td>
<td></td>
</tr>
<tr>
<td></td>
<td>2. Lack of resources for transition programs</td>
<td>2. S</td>
<td></td>
</tr>
<tr>
<td>Adult Heme RNs (N = 1)</td>
<td>1. Patients are required to have a primary physician to manage their medical care</td>
<td>1. S</td>
<td>Only one individual responded, barriers as indicated.</td>
</tr>
<tr>
<td></td>
<td>2. Patients are not responsible for their care</td>
<td>2. I</td>
<td></td>
</tr>
<tr>
<td></td>
<td>3. No social worker and/or nurse practitioner</td>
<td>3. S</td>
<td></td>
</tr>
<tr>
<td></td>
<td>4. No staff dedicated to patients with SCD</td>
<td>4. S</td>
<td></td>
</tr>
<tr>
<td>Pediatric H/O RNs (N = 6)</td>
<td>1. Loss of insurance</td>
<td>1. S</td>
<td>The most frequently expressed barriers were the lack of adult providers, lack of information about transition provided to patients and patients reluctance to leave pediatrics because of the trust that they have developed with their HC providers</td>
</tr>
<tr>
<td></td>
<td>2. Patients and families do not want to leave</td>
<td>2. P &amp; I</td>
<td></td>
</tr>
<tr>
<td></td>
<td>3. MDs are not ready to transition</td>
<td>3. P</td>
<td></td>
</tr>
<tr>
<td></td>
<td>4. Lack of adult providers</td>
<td>4. P &amp; S</td>
<td></td>
</tr>
<tr>
<td></td>
<td>5. Maturity level of patients</td>
<td>5. I</td>
<td></td>
</tr>
<tr>
<td></td>
<td>6. Reluctance of adult providers to accept patients</td>
<td>6. P</td>
<td></td>
</tr>
<tr>
<td></td>
<td>7. Delayed or no preparation for transitioning</td>
<td>7. P</td>
<td></td>
</tr>
<tr>
<td></td>
<td>8. Patient/Physician trust</td>
<td>8. I</td>
<td></td>
</tr>
<tr>
<td>Emergency Dept</td>
<td></td>
<td></td>
<td>Fear of leaving pediatrics and leaving their pediatric health care provider were the most commonly identified barriers. Lack of adult providers, lack of resources to support transition and failure to prepare young adults for adult health care services were also barriers</td>
</tr>
<tr>
<td>Peds RNs (N = 13)</td>
<td>1. Fear of leaving pediatrics</td>
<td>1. I</td>
<td></td>
</tr>
<tr>
<td></td>
<td>2. Patients do not want to leave their pediatric health care provider</td>
<td>2. I</td>
<td></td>
</tr>
<tr>
<td></td>
<td>3. Inability of providers to prepare patients and families for transitioning to adult care</td>
<td>3. P &amp; S</td>
<td></td>
</tr>
<tr>
<td></td>
<td>4. Lack of adult providers</td>
<td>4. P &amp; S</td>
<td></td>
</tr>
<tr>
<td></td>
<td>5. Lack of resources to support transition</td>
<td>5. S</td>
<td></td>
</tr>
</tbody>
</table>
| Pediatric H/O NPs (N = 4) | 1. Lack of adult providers  
2. Loss of medical home  
3. Pediatric providers do not want to transition  
4. Patients are not prepared to transition  
5. Patients and families do not want to leave pediatrics  
6. Location of adult clinic in another building  
7. Adult providers reluctance to prescribe pain medication when in crisis  
8. Fear of the unknown  
9. Perception that young adult is a child  
10. Intellectual deficits as a result of stroke  
11. Lack of knowledge about selfcare (making appointments and medication calling for refills) | 1. P & S  
2. S  
3. P  
4. P  
5. I  
6. S  
7. P & S  
8. I  
9. P  
10. I  
11. I | All participants expressed the lack of adult providers as a major barrier. Health care services in pediatrics is organized around a comprehensive, family-centered model while adult care is more individualized. Preparation of the young adult to assume selfcare responsibilities after they transition to adult services, location of services in a different building, as well as pediatric providers reluctance to refer to adult providers. |
| --- | --- | --- |
| Pediatric H/O MD/DO (N = 5) | 1. Different model of care in pediatric and medical subspecialties  
2. Lack of medical home  
3. Lack of providers for young adults  
4. Lack of interest from medical providers to provide and coordinate care  
5. Pediatric doctors don’t push patients  
6. A formal process  
7. No financial support/lack of resources | 1. S  
2. S  
3. S & P  
4. S & P  
5. S & P  
6. P  
7. S | The most frequently expressed barrier was the lack of adult providers to take young adults with complex medical conditions like SCD. |