Disease Knowledge and Readiness for Transition in Adolescents with Sickle Cell Disease in Jamaica: A Mixed-Methods Study

by

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Thesis submitted in partial fulfillment of
the requirements for the degree of
Master of Science in the Global Health in the Duke Global Health Institute
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ABSTRACT

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Abstract

Introduction: Sickle cell disease (SCD) is a genetically inherited recessive blood disorder that affects millions of people worldwide. The management of SCD should and can be considered a collaborative team effort, and requires the comprehensive and coordinated support of several medical professionals. The rising number of adults living with SCD creates a need for long term therapeutic and management strategies as well as a better understanding of the transition from pediatric to adult care. The research goal for this project is to compare the two systems that exist for treatment of adolescents with SCD in Jamaica and the United States by assessing differences and similarities in patients' readiness for adult treatment and their understanding of SCD and its management. Methods: This study was conducted in the Sickle Cell Unit at the University of West Indies (UWI) hospital in Kingston, Jamaica. Eligibility for this study was defined as patients with SCD, between the ages of 13-19, seeking treatment at the health facility in the University of the West Indies, who have no acute illness at the time of study. After a verbal and written consent process during check-in, each participant completed a demographic survey, disease knowledge questionnaire, the ASH Transition Readiness Assessment Questionnaire, and had the opportunity to participate in in-depth interviews. Following data collection, results subsequently with similar previously completed surveys from patients at the Duke University Sickle Cell Center. Results:

Gender and socioeconomic factors were not associated with differences in assessment scores in Jamaica. Total scores for disease knowledge questionnaires increased with age, however mean scores for the 17-19 age group were 62.17% lower than Duke University patients of the same age. Self-evaluation with the ASH Transition Readiness Assessment also showed an increase in scores with age, and significant increases in *disease knowledge* and *appointments* sections in both the 13-14 and 17-19 age groups, estimated by a p-value of 0.023 and 0.006, respectively. The results, however, were also generally lower than similar Transition Readiness Assessment measures at Duke. In-depth interviews revealed patient insight into disease knowledge, treatment involvement and experiences with doctors, family, and in the clinic. Answers align with both questionnaires used in this study.

Dedication

This thesis is dedicated to the wonderful young people who took part in this study and are contributing to the understanding of sickle cell disease transition in Jamaica and beyond.

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Acknowledgements

Thank you to the Duke Global Health Institute, the Sickle Cell Unit, Caribbean Institute for Health Research of the University of the West Indies, and Duke University Sickle Cell Center for all of their help, guidance, and support.

Thank you to the wonderful nurses and volunteers who accepted me as one of their own.

Thank you to my husband, parents and younger sister for their unconditional love, strength, and support.

1. Introduction

Sickle cell disease (SCD) is a genetically inherited recessive blood disorder that affects millions of people worldwide. First reported in 1910 by James Herrick and his intern Ernest Irons who were managing a severely anemic Chicago dental student from Grenada, it was classified as the first molecular disease in 1945.[1] The condition is caused by a point mutation in the beta (β) subunit of the hemoglobin gene that converts valine to glutamic acid.[2, 3] One copy renders a person a carrier, while two copies of this mutation lead to molecular changes in deoxygenated erythrocytes that result in loss of flexibility, giving way to the characteristic "sickle" shape. The sickling of erythrocytes gives rise to the most defining symptom: pain. Vaso-occlusive crisis occurs when cells that have become sickled block blood vessels, leading to intense pain and inflammation.[4] No two patients with SCD, however, are the same due to a high level of phenotypic variability.[5] For some, the pain associated with vaso-occlusive crisis is incapacitating while others have milder symptoms. Treatment of SCD usually involves the use of

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[1] McGann, Patrick T, Nero, Alecia C, and Ware, Russel E. 2017. "Clinical Features of β-thalassemia and Sickle Cell Disease" *Springer Link* 1-26.

[2] Stuart, Marie J., and Ronald L. Nagel. 2004. "Sickle-Cell Disease." *Lancet (London, England)* 364 (9442): 1343–60.

[3] Ware, Russell E, de Montalembert, Mariane, Léon Thsilolo, and Abboud, Miguel R. 2017. "Sickle Cell Disease " *Lancet* 390 (10091): 311-23.

[4] Stuart, Marie J., and Ronald L. Nagel. 2004. "Sickle-Cell Disease." *Lancet (London, England)* 364 (9442): 1343–60.

[5] Thein, Swee Lay. 2017. "Genetic Basis and Genetic Modifiers of β-Thalassemia and Sickle Cell Disease." *SpringerLink*, 27–57.

opiates, leading to societal and cultural stigma around patients, under-treatment for pain, labelling them as drug seekers, as well as complicated relationships between patients and providers.[6] Funding for SCD has been substantially lower than other monogenic diseases. This limitation directly affects new innovations in research, therapies, and clinical care, which in turn affect quality of health care and outcomes for SCD patients.[7]

1.1 Disease Pathophysiology

Within erythrocytes, hemoglobin is the oxygen carrying component which bind to and carries oxygen from the lungs to tissues, and carbon dioxide from tissues to lungs. It consists of 4 globin chains, two alpha (α) and two beta (β) subunits, with an iron heme group.[8] Classified as a monogenic hemoglobinopathy, which includes thalassemias as well as sickle cell syndromes, these conditions are the most common Mendelian genetic diseases in the world.[9, 10]

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[6] Stuart, Marie J., and Ronald L. Nagel. 2004. "Sickle-Cell Disease." *Lancet (London, England)* 364 (9442): 1343–60.

[7] Glassberg, Jeffrey, and Michael R. DeBaun. 2017. "Sickle Cell Disease." *SpringerLink*, 131–38.

[8] Ekong, Dr Anietie. 2017. "Sickle Cell Disease." *InnovAiT* 10 (2): 73–81.

[9] Thein, Swee Lay. 2017. "Genetic Basis and Genetic Modifiers of β-Thalassemia and Sickle Cell Disease." *SpringerLink*, 27–57.

[10] Ekong, Dr Anietie. 2017. "Sickle Cell Disease." *InnovAiT* 10 (2): 73–81.

The term sickle cell disease, does not refer to only one disease manifestation but rather to a collection of inherited blood disorders.[11] β-thalassemia and SCD are the most clinically significant, and while SCD is caused by a single nucleotide substitution on the sixth codon of the HBB gene from adenine to thymine (βGlu6Val), β-thalassemia is due to an array of mutations causing structural changes to the β-globin subunit.[12, 13] HbS, the sickle variant of normal adult hemoglobin, denoted HbA, is inherited from one or both parents and can be inherited alongside other variants such as HbC or β-thalassemias.[14, 15]

Symptoms typically do not arise until 6-12 months, due to elevated and persistent levels of HbF hemoglobin in infants that begin to taper off with age.[16] SCD is a disorder that eventually affects nearly every organ in the body. When the hemoglobin becomes deoxygenated, HbS polymerizes, losing cations and water and taking on abnormal structural changes unfavorable for movement throughout blood vessels,

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[11] Ware, Russell E, de Montalembert, Mariane, Léon Thsilolo, and Abboud, Miguel R. 2017. "Sickle Cell Disease " *Lancet* 390 (10091): 311-23.

[12] Thein, Swee Lay. 2017. "Genetic Basis and Genetic Modifiers of β-Thalassemia and Sickle Cell Disease." *SpringerLink*, 27–57.

[13] Ware, Russell E, de Montalembert, Mariane, Léon Thsilolo, and Abboud, Miguel R. 2017. "Sickle Cell Disease " *Lancet* 390 (10091): 311-23.

[14] Piel, Frédéric B., Martin H. Steinberg, and David C. Rees. 2017. "Sickle Cell Disease." *New England Journal of Medicine* 376 (16): 1561–73.

[15] Ware, Russell E, de Montalembert, Mariane, Léon Thsilolo, and Abboud, Miguel R. 2017. "Sickle Cell Disease " *Lancet* 390 (10091): 311-23.

[16] Cheesman, Simon. 2015. "Sickle Cell Disease: Symptoms, Complications and Management." Pharmaceutical Journal. Accessed February 9, 2018. https://www.pharmaceutical-journal.com/learning/learning-article/sickle-cell-disease-symptoms-complications-and-management/20069268.article.

resulting in hemolytic anemia, vaso-occlusive crisis and ischemic damage to tissues.[17,18] The cascade of events from vaso-occlusion and the complications that arise are central to what characterizes SCD. Chronic complications can either be related to the affected large blood vessels or by progressive organ damage.[19] Among the severe complications are acute chest syndrome, stroke, pulmonary hypertension, renal damage, retinopathies, and leg ulcers.[20]

Compound heterozygosity of SCD, or the inheritance of HbS with another variant, is common among patients and results in either more or less severe clinical implications. However, each genotype still exhibits phenotypic variability among patients. Expression of HbF (fetal hemoglobin) and α-thalassemia may modify SCD presentation.[21] Additionally, studies have shown that HbS haplotypes can also affect phenotypic expression.[22] Further research needs to be done to understand this wide

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[17] Piel, Frédéric B., Martin H. Steinberg, and David C. Rees. 2017. "Sickle Cell Disease." *New England Journal of Medicine* 376 (16): 1561–73.

[18] Stuart, Marie J., and Ronald L. Nagel. 2004. "Sickle-Cell Disease." *Lancet (London, England)* 364 (9442): 1343–60.

[19] Piel, Frédéric B., Martin H. Steinberg, and David C. Rees. 2017. "Sickle Cell Disease." *New England Journal of Medicine* 376 (16): 1561–73.

[20] Cheesman, Simon. 2015. "Sickle Cell Disease: Symptoms, Complications and Management." Pharmaceutical Journal. Accessed February 9, 2018. https://www.pharmaceutical-journal.com/learning/learning-article/sickle-cell-disease-symptoms-complications-and-management/20069268.article.

[21] Piel, Frédéric B., Martin H. Steinberg, and David C. Rees. 2017. "Sickle Cell Disease." *New England Journal of Medicine* 376 (16): 1561–73.

[22] Tyghter, S. Staniforth, Ware, M., D. and G. Serjeant. 1998. "Airline Travel in Sickle-Cell Disease." *Lancet (London, England)* 352 (9128): 652.

range of patient experiences with SCD. Implications for this could drastically improve the standard of care worldwide.

1.2 Types of Sickle Cell Disease

Hemoglobinopathies, like sickle cell disorders, are conditions that affect the structure of hemoglobin and encompass the various monogenic, inherited, mutations that occur in several parts of the hemoglobin molecule in people all over the world. Some are more prevalent among certain racial/ethnic groups than others. Interestingly, clinical ramifications within the same genotype can also vary by individual. Sickle cell *anemia* (SCA) refers to inheritance of two copies of the HbS allele, denoted HbSS, while SCD includes SCA and conditions where one HbS allele and another hemoglobin allele are inherited.[23] There are several types of hemoglobinopathies, but three have been highlighted here for purposes of this study.

1.2.1 Type HbSC

Occurring when one HbS allele is inherited with the HbC allele, sickle cell HbSC accounts for 30-35% of the SCD population of African descent.[24] This genotype usually presents more moderate symptoms and expression of HbS and HbC are around 50%

[23] Thein, Swee Lay. 2017. "Genetic Basis and Genetic Modifiers of β-Thalassemia and Sickle Cell Disease." *SpringerLink*, 27–57.
[24] Thein, Swee Lay. 2017. "Genetic Basis and Genetic Modifiers of β-Thalassemia and Sickle Cell Disease." *SpringerLink*, 27–57.

and 45% respectively after laboratory electrophoresis.[25] As with all SCD genotypes, phenotypic variation among individuals is along a spectrum. However, patients with HbSC are more prone to proliferative sickle retinopathy (PSR) and splenomegaly.[26] A common complication with SCD in children, splenomegaly in childhood can persist into adulthood leading to a risk of acute splenic sequestration crisis.[27]

1.2.2 Type HbS-beta thalassemia

Overall, there have been more than 300 β-thalassemias identified, but only 40 account for 90% of all thalassemias worldwide. Associated with SCD are HbS β^+ and HbS β^0 where compound heterozygosity is exhibited through inheriting both the HbS allele and β-thalassemia.[28] This type of SCD exhibits a wide clinical spectrum that depends on the kind of mutation, and the amount of normal hemoglobin, HbA, or HbF produced.[29] For HbS β^0, there is no production of HbA and is almost indistinguishable from HbSS with symptoms generally being more severe. In individuals with HbS β^+, HbA production can vary between 15-25% and can range from 5-30%. HbF production

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[25] McGann, Patrick T, Nero, Alecia C, and Ware, Russel E. 2017. "Clinical Features of β-thalassemia and Sickle Cell Disease" *Springer Link* 1-26.

[26] Serjeant, Graham R. 2013. "The Natural History of Sickle Cell Disease." *Cold Spring Harbor Perspectives in Medicine* 3 (10)..

[27] McGann, Patrick T, Nero, Alecia C, and Ware, Russel E. 2017. "Clinical Features of β-thalassemia and Sickle Cell Disease" *Springer Link* 1-26.

[28] Thein, Swee Lay. 2017. "Genetic Basis and Genetic Modifiers of β-Thalassemia and Sickle Cell Disease." *SpringerLink*, 27–57.

[29] Serjeant, Graham R. 2013. "The Natural History of Sickle Cell Disease." *Cold Spring Harbor Perspectives in Medicine* 3 (10).

can be anywhere from 1-20%.[30,31] Variability in hemoglobin expression contributes to differences in phenotypic presentation. This type of SCD is usually mild among African patients, but severe among those of Mediterranean, Asian, or Indian decent (Thein, 2017).[32] In fact, HbS β^+ in Indians only produce 3-5% HbA with this specific genotype.[33]

1.2.3 Type HbSS

The most clinically severe genotype, HbSS, is the result of inheriting two HbS alleles. HbS arose independently in 4 locations, 3 originating in Sub-Saharan Africa and one in the Arab peninsula and Indian subcontinent.[34] Moreover, HbSS may account for up to 65-70% of all SCD in African populations.[35] The varying severity of HbSS among different individuals and racial and ethnic populations is still not completely understood, but may be attributed to the genetic polymorphisms that surround the HbS mutation, creating differences in HbF expression. Senegalese and Arabs have genetic polymorphisms that lead to higher circulating HbF and relatively milder symptoms.

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[30] Thein, Swee Lay. 2017. "Genetic Basis and Genetic Modifiers of β-Thalassemia and Sickle Cell Disease." *SpringerLink*, 27–57.

[31] Serjeant, Graham R. 2013. "The Natural History of Sickle Cell Disease." *Cold Spring Harbor Perspectives in Medicine* 3 (10).

[32] Thein, Swee Lay. 2017. "Genetic Basis and Genetic Modifiers of β-Thalassemia and Sickle Cell Disease." *SpringerLink*, 27–57.

[33] Serjeant, Graham R. 2013. "The Natural History of Sickle Cell Disease." *Cold Spring Harbor Perspectives in Medicine* 3 (10).

[34] McGann, Patrick T, Nero, Alecia C, and Ware, Russel E. 2017. "Clinical Features of β-thalassemia and Sickle Cell Disease" *Springer Link* 1-26.

[35] Thein, Swee Lay. 2017. "Genetic Basis and Genetic Modifiers of β-Thalassemia and Sickle Cell Disease." *SpringerLink*, 27–57.

Meanwhile, symptoms in those from Central Africa are more severe as their polymorphisms lead to less HbF expression.[36]

1.3 Burden of Disease

SCD prevalence is high in certain regions of Africa, the Mediterranean/Arabian Peninsula, India, and the Caribbean.[37, 38] An individual who is heterozygous for the HbS gene has a heightened level of protection against malaria, giving the gene a selective advantage and ensuring its survival into subsequent generations.[39] The Global Burden of disease estimated that the prevalence of the sickle cell trait increased from 338,756 to 404,566 between 2005 and 2015, a 19.4% change in 10 years.[40] In the United States, 100,000 people have SCD. Though it is difficult to estimate globally, approximately 300,000 babies are born each year are born with SCD and 5.4 million are born with the

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[36] McGann, Patrick T, Nero, Alecia C, and Ware, Russel E. 2017. "Clinical Features of β-thalassemia and Sickle Cell Disease" *Springer Link* 1-26.

[37] Piel, Frédéric B., Martin H. Steinberg, and David C. Rees. 2017. "Sickle Cell Disease." *New England Journal of Medicine* 376 (16): 1561–73.

[38] Stuart, Marie J., and Ronald L. Nagel. 2004. "Sickle-Cell Disease." *Lancet (London, England)* 364 (9442): 1343–60.

[39] Piel, Frédéric B., Martin H. Steinberg, and David C. Rees. 2017. "Sickle Cell Disease." *New England Journal of Medicine* 376 (16): 1561–73.

[40] "Global, Regional, and National Incidence, Prevalence, and Years Lived with Disability for 310 Diseases and Injuries, 1990–2015: A Systematic Analysis for the Global Burden of Disease Study 2015." 2016. *The Lancet* 388 (10053): 1545–1602.

trait.[41, 42] Additionally, it is estimated that Nigeria, India, and the Democratic Republic of Congo share half the global burden of 300,000 newborns.[43, 44] Overall, approximately 70% of cases occur in people of African descent with a prevalence of 2-30% of the trait in African populations.[45, 46] It is estimated that by the year 2050, the number of affected births could increase by 30% with the expected population increase.[47]

The number of people living with SCD is expected to increase in both high income and low income countries, due to increasing life expectancy and advances in medical technologies. In fact, over the past 40 years, interventions in high income countries have leveled out childhood mortality with the general population. Although childhood mortality for individuals with SCD has decreased in low income countries, it can reach up to 90% in low resource settings lacking the infrastructure for basic health care.[48]

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[41] Shet, Arun S., and Swee Lay Thein. 2017. "Therapeutic Advances in Sickle Cell Disease in the Last Decade." *The Indian Journal of Medical Research* 145 (6): 708.

[42] Ware, Russell E, de Montalembert, Mariane, Léon Thsilolo, and Abboud, Miguel R. 2017. "Sickle Cell Disease " *Lancet* 390 (10091): 311-23.

[43] Piel, Frédéric B., Martin H. Steinberg, and David C. Rees. 2017. "Sickle Cell Disease." *New England Journal of Medicine* 376 (16): 1561–73.

[44] Shet, Arun S., and Swee Lay Thein. 2017. "Therapeutic Advances in Sickle Cell Disease in the Last Decade." *The Indian Journal of Medical Research* 145 (6): 708.

[45] Stuart, Marie J., and Ronald L. Nagel. 2004. "Sickle-Cell Disease." *Lancet (London, England)* 364 (9442): 1343–60.

[46] Ware, Russell E, de Montalembert, Mariane, Léon Thsilolo, and Abboud, Miguel R. 2017. "Sickle Cell Disease " *Lancet* 390 (10091): 311-23.

[47] Ware, Russell E, de Montalembert, Mariane, Léon Thsilolo, and Abboud, Miguel R. 2017. "Sickle Cell Disease " *Lancet* 390 (10091): 311-23.

[48] Piel, Frédéric B., Martin H. Steinberg, and David C. Rees. 2017. "Sickle Cell Disease." *New England Journal of Medicine* 376 (16): 1561–73.

Overall, thalassemias, SCD, and their traits still heavily contribute to years of life lost (YLLs) and over 100,000 deaths, a 6% increase from 2005 to 2015, due to their associated complications.[49] Thalassemias and the trait contribute 31,300 and 3.92 million YLLs respectively, while SCD and the trait contributed to 371,400 and 1.72 million YLLs respectively during the same 10 year span.[50] The economic burden of SCD is important, particularly in low resource areas where burden is highest, and the need for cost-effective screening and interventions would radically reduce the 15% of total childhood mortality under five that is attributable to SCD.[51] In 2008, the United Nations named SCD a global health priority, which led to a WHO supplemental report that provided specific targets and goals for sub-Saharan countries. However, despite high burden in Africa, tens of thousands of births occur in the Americas and Middle East each year, and populations are emerging each year in countries where previously SCD prevalence was negligible.[52]

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[49] Ware, Russell E, de Montalembert, Mariane, Léon Thsilolo, and Abboud, Miguel R. 2017. "Sickle Cell Disease " *Lancet* 390 (10091): 311-23.

[50] "Global, Regional, and National Incidence, Prevalence, and Years Lived with Disability for 310 Diseases and Injuries, 1990–2015: A Systematic Analysis for the Global Burden of Disease Study 2015." 2016. *The Lancet* 388 (10053): 1545–1602.

[51] Ware, Russell E, de Montalembert, Mariane, Léon Thsilolo, and Abboud, Miguel R. 2017. "Sickle Cell Disease " *Lancet* 390 (10091): 311-23.

[52] Ware, Russell E. 2013. "Is Sickle Cell Anemia a Neglected Tropical Disease?" *PLoS Neglected Tropical Diseases* 7 (5).

1.4 Treatment of Sickle Cell Disease

The management of SCD should and can be considered a collaborative team effort, and requires the comprehensive and coordinated support of several medical professionals. These include a hematology team and specialities with experience dealing with SCD complications such as pulmonology, neurology, ophthalmology, orthopedic, renal and pre-natal care.[53] Chronic pain in individuals with SCD can result in poor quality of life. The co-morbidities that are associated with pain are depression, anxiety, insomnia, and dependence on pain medications, making access to psychological and physical therapies, social services, and local and religious support groups an essential part of sickle cell care.[54, 55] Along with supportive care, over the past few decades, there have been several medical advancements being used in practice today, and emerging studies in gene therapy show promise to correct the physiological malfunction at the source.

1.4.1 Current Treatments

The two most widely used therapies to treat chronic symptoms of SCD are hydroxyurea and blood transfusions. Hydroxyurea, also known as hydroxycarbamide,

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[53] Stuart, Marie J., and Ronald L. Nagel. 2004. "Sickle-Cell Disease." *Lancet (London, England)* 364 (9442): 1343–60.

[54] Yawn, Barbara P., George R. Buchanan, Araba N. Afenyi-Annan, Samir K. Ballas, Kathryn L. Hassell, Andra H. James, Lanetta Jordan, et al. 2014. "Management of Sickle Cell Disease: Summary of the 2014 Evidence-Based Report by Expert Panel Members." *JAMA* 312 (10): 1033.

[55] Stuart, Marie J., and Ronald L. Nagel. 2004. "Sickle-Cell Disease." *Lancet (London, England)* 364 (9442): 1343–60.

when taken orally on a daily basis has been shown to reduce acute and chronic events.[56] It has been shown to be safe in both children and adults, yet continues to be underused due to high costs, few resources and lack of experience for monitoring and dosing[57, 58, 59] Identified in the 1980s, hydroxyurea works by increasing the expression of HbF in erythrocytes, preventing them from sickling.[60, 61] It also reduces the number of leukocytes and reticulocytes circulating in the blood stream, increases red blood cell size and deformability, and may contribute to vasodilation through the release of nitric oxide during the metabolism of the drug. In total, long term hydroxyurea therapy has shown a reduction in pain crises, hospitalizations, and mortality in patients.[62] Until 2017, hydroxyurea was the only FDA approved treatment. However, a new drug named

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[56] Yawn, Barbara P., George R. Buchanan, Araba N. Afenyi-Annan, Samir K. Ballas, Kathryn L. Hassell, Andra H. James, Lanetta Jordan, et al. 2014. "Management of Sickle Cell Disease: Summary of the 2014 Evidence-Based Report by Expert Panel Members." *JAMA* 312 (10): 1033.

[57] Cheesman, Simon. 2015. "Sickle Cell Disease: Symptoms, Complications and Management." Pharmaceutical Journal. Accessed February 9, 2018. https://www.pharmaceutical-journal.com/learning/learning-article/sickle-cell-disease-symptoms-complications-and-management/20069268.article.

[58] Piel, Frédéric B., Martin H. Steinberg, and David C. Rees. 2017. "Sickle Cell Disease." *New England Journal of Medicine* 376 (16): 1561–73.

[59] Ware, Russell E, de Montalembert, Mariane, Léon Thsilolo, and Abboud, Miguel R. 2017. "Sickle Cell Disease " *Lancet* 390 (10091): 311-23.

[60] Cheesman, Simon. 2015. "Sickle Cell Disease: Symptoms, Complications and Management." Pharmaceutical Journal. Accessed February 9, 2018. https://www.pharmaceutical-journal.com/learning/learning-article/sickle-cell-disease-symptoms-complications-and-management/20069268.article.

[61] Yawn, Barbara P., George R. Buchanan, Araba N. Afenyi-Annan, Samir K. Ballas, Kathryn L. Hassell, Andra H. James, Lanetta Jordan, et al. 2014. "Management of Sickle Cell Disease: Summary of the 2014 Evidence-Based Report by Expert Panel Members." *JAMA* 312 (10): 1033.

[62] Yawn, Barbara P., George R. Buchanan, Araba N. Afenyi-Annan, Samir K. Ballas, Kathryn L. Hassell, Andra H. James, Lanetta Jordan, et al. 2014. "Management of Sickle Cell Disease: Summary of the 2014 Evidence-Based Report by Expert Panel Members." *JAMA* 312 (10): 1033..

Endari, an L-glutamine oral powder has now been approved for use and has shown effective results in both adults and children under 5.

Blood transfusions in patients with SCD serve two main purposes, to address anemia and to lower the risk of stroke from repeated vaso-occlusive crises by replacing abnormal red blood cells with donor blood expressing the normal HbA allele. To address, anemia, transfusions occur episodically. In order to prevent iron overload, the patient's blood is exchanged with donor blood. Patients who receive several blood transfusions may need iron chelation to mitigate these effects. Other complications include, hyperviscosity of the blood and hemolysis. [63, 64]

Stem cell therapy through HLA-matched sibling cord blood or bone marrow transplantation is the only viable option for a cure in patients with SCD as gene transfer is possible, but precarious. However, access to this is severely limited, as only 10-20% patients have siblings who concurrently do not have SCD and are matches to patients. Opening donor pools to HLA-matched unrelated individuals has not been successful, further restricting accessibility. [65] Bone marrow transplantation can help ameliorate

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[63] Cheesman, Simon. 2015. "Sickle Cell Disease: Symptoms, Complications and Management." Pharmaceutical Journal. Accessed February 9, 2018. https://www.pharmaceutical-journal.com/learning/learning-article/sickle-cell-disease-symptoms-complications-and-management/20069268.article.

[64] Yawn, Barbara P., George R. Buchanan, Araba N. Afenyi-Annan, Samir K. Ballas, Kathryn L. Hassell, Andra H. James, Lanetta Jordan, et al. 2014. "Management of Sickle Cell Disease: Summary of the 2014 Evidence-Based Report by Expert Panel Members." *JAMA* 312 (10): 1033.

[65] Ware, Russell E, de Montalembert, Mariane, Léon Thsilolo, and Abboud, Miguel R. 2017. "Sickle Cell Disease " *Lancet* 390 (10091): 311-23.

symptoms, but is incredibly invasive and related to an extensive list of complications, which makes the need for further research on therapeutic options vital to those affected. Several pathways in the pathophysiology of the disease can be targets for new medical therapies, including interruption of vaso-occlusion and inhibition of HbS polymerization.[66, 67, 68]

1.4.2 Potential Future Treatments

Innovations in SCD treatment are being made, but pathways have yet to be fully explored. New compounds in clinical trials, mainly focusing adhesion and inflammation by targeting of selectins in erythrocytes, leukocytes, platelets, and the endothelium.[69, 70] Studies involving gene therapy have been successful in mice and recently in one patient in France.[71] Two studies involved editing hematopoietic stem cells to induce the expression or "de-repression" of HbF and then transplanted into the marrow. In one study, genes were altered through a lentiviral vector and long term expression was

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[66] Yawn, Barbara P., George R. Buchanan, Araba N. Afenyi-Annan, Samir K. Ballas, Kathryn L. Hassell, Andra H. James, Lanetta Jordan, et al. 2014. "Management of Sickle Cell Disease: Summary of the 2014 Evidence-Based Report by Expert Panel Members." *JAMA* 312 (10): 1033.

[67] Shet, Arun S., and Swee Lay Thein. 2017. "Therapeutic Advances in Sickle Cell Disease in the Last Decade." *The Indian Journal of Medical Research* 145 (6): 708.

[68] Ware, Russell E, de Montalembert, Mariane, Léon Thsilolo, and Abboud, Miguel R "Sickle Cell Disease." 2017. *The Lancet* 390 (10091): 311–23.

[69] Stuart, Marie J., and Ronald L. Nagel. 2004. "Sickle-Cell Disease." *Lancet (London, England)* 364 (9442): 1343–60.

[70] Ware, Russell E, de Montalembert, Mariane, Léon Thsilolo, and Abboud, Miguel R "Sickle Cell Disease." 2017. *The Lancet* 390 (10091): 311–23.

[71] Ribeil, Jean-Antoine, et al. 2017. "Gene Therapy in a Patient with Sickle Cell Disease" *The New England Journal of Medicine* (376): 848-855.

achieved and found HbF in 52% of circulating cells.[72, 73] More recently a newer technology, CRISPR/Cas9, has been shown as an effective strategy to mitigate some of the negative aspects of lentiviral vectors. Results from this study show an increase in circulating erythrocytes expressing HbF up to 39%.[74]

1.5 Transition from Pediatric Care to Adult Care

With advances in SCD treatment, patients are now living longer and survive into adulthood, which was not usually possible 30 years ago, even more so in high-resource settings.[75, 76] This is due to improved newborn screening, penicillin prophylaxis, pneumococcal immunizations, and education.[77, 78] The rising number of adults living with SCD creates a need for long term therapeutic and management strategies as well as

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[72] Stuart, Marie J., and Ronald L. Nagel. 2004. "Sickle-Cell Disease." *Lancet (London, England)* 364 (9442): 1343–60.

[73] Pawliuk, R., K. A. Westerman, M. E. Fabry, E. Payen, R. Tighe, E. E. Bouhassira, S. A. Acharya, et al. 2001. "Correction of Sickle Cell Disease in Transgenic Mouse Models by Gene Therapy." *Science (New York, N.Y.)* 294 (5550): 2368–71.

[74] Lin, Michelle I., Elizabeth Paik, Bibhu Mishra, David Burkhardt, Andrew Kernytsky, Michael Pettiglio, Yi-Shan Chen, et al. 2017. "CRISPR/Cas9 Genome Editing to Treat Sickle Cell Disease and B-Thalassemia: Re-Creating Genetic Variants to Upregulate Fetal Hemoglobin Appear Well-Tolerated, Effective and Durable." *Blood* 130 (Suppl 1): 284–284.

[75] Andemariam Biree, Owarish-Gross Jasmine, Grady James, Boruchov Donna, Thrall Roger S., and Hagstrom J. Nathan. 2014. "Identification of Risk Factors for an Unsuccessful Transition from Pediatric to Adult Sickle Cell Disease Care." *Pediatric Blood & Cancer* 61 (4): 697–701.

[76] Frost, Jennifer R., Rebecca K. Cherry, Suzette O. Oyeku, Elissa Z. Faro, Lori E. Crosby, Maria Britto, Lisa K. Tuchman, Ivor B. Horn, Charles J. Homer, and Anjali Jain. 2016. "Improving Sickle Cell Transitions of Care Through Health Information Technology." *American Journal of Preventive Medicine*, Developing a Unified Approach for Sickle Cell Disease, 51 (1, Supplement 1): S17–23.

[77] Ware, Russell E, de Montalembert, Mariane, Léon Thsilolo, and Abboud, Miguel R "Sickle Cell Disease." 2017. *The Lancet* 390 (10091): 311–23.

[78] Ware, Russell E. 2013. "Is Sickle Cell Anemia a Neglected Tropical Disease?" *PLoS Neglected Tropical Diseases* 7 (5).

a better understanding of the transition from pediatric to adult care. Efforts have been made to address this need in the United States; however, SCD is a global disease.

Naturally, what comprises transition and whether a framework is present depends on the region where care is provided. Barriers to successful transition include: the patients and their families being overly dependent on pediatric providers, the pediatric providers fostering dependency in their patients, and a lack of communication between pediatric and adult providers.[79, 80] Moreover, successful transition is paramount to improvments in survival. It was found in the Dallas Cohort Study that patients are at greatest risk for mortality when they are transition aged, caused primarily by acute chest syndrome, stroke, and organ failure.[81] There are no statistics on similar adolescent populations in Jamaica and limited information on other countries SCD transition should be a long-term process that gradually prepares the patients for responsibility, management and independence, and should not be defined as a discrete event.[82]

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[79] Ware, Russell E. 2013. "Is Sickle Cell Anemia a Neglected Tropical Disease?" *PLoS Neglected Tropical Diseases* 7 (5).

[80] Telfair, Joseph, Leah R. Alexander, Penny S. Loosier, Patty L. Alleman-Velez, and Julie Simmons. 2004. "Providers' Perspectives and Beliefs Regarding Transition to Adult Care for Adolescents with Sickle Cell Disease." *Journal of Health Care for the Poor and Underserved* 15 (3): 443–61.

[81] Andemariam Biree, Owarish-Gross Jasmine, Grady James, Boruchov Donna, Thrall Roger S., and Hagstrom J. Nathan. 2014. "Identification of Risk Factors for an Unsuccessful Transition from Pediatric to Adult Sickle Cell Disease Care." *Pediatric Blood & Cancer* 61 (4): 697–701.

[82] Frost, Jennifer R., Rebecca K. Cherry, Suzette O. Oyeku, Elissa Z. Faro, Lori E. Crosby, Maria Britto, Lisa K. Tuchman, Ivor B. Horn, Charles J. Homer, and Anjali Jain. 2016. "Improving Sickle Cell Transitions of Care Through Health Information Technology." *American Journal of Preventive Medicine*, Developing a Unified Approach for Sickle Cell Disease, 51 (1, Supplement 1): S17–23.

Achieving a high quality standard of care for this poorly understood niche of treatment is of the utmost importance, with the need growing every day.

Center for Healthcare Transition and Improvement, or Got Transition, is a collaboration between maternal and child health bureau and the national alliance to advance adolescent health. Their mission is to improve transition from pediatric to adult care through use of new and innovative strategies. They outline six core elements to health care transition, as well as highlight cultural competence: Transition policy, transition tracking and monitoring, transition readiness, transition planning, transfer of care, transfer completion. They were also involved in the release of a general Transition Readiness Assessment Questionnaire (TRAQ) for a variety of medical conditions.[83] Additionally, an international Delphi study interviewed providers on what they believed were key elements and indicators for successful transition. The study found an agreement on six essential key elements, and one indicator: Coordination between providers, planning transition at an early age, self-management, incorporating youth's perspectives and preferences, attending appointments alone, and identifying an adult provider. The indicator was patient loss to follow up. Findings for this study could be used as a framework to develop guidelines at a local, national, and international level, however had a heavily western perspective. For purposes of this study, there are no known policies for transition in Jamaica.[84]

Frameworks for SCD treatment differ globally and infrastructure for healthcare depends on the environment and resources available. Two examples of unique

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[83] GotTransition.org. n.d. "GotTransition.org - About Got Transition." GotTransition.org. Accessed March 17, 2018. http://www.gottransition.org/about/index.cfm.

[84] Suris, Joan-Carles, and Christina Akre. 2015. "Key Elements For, and Indicators Of, a Successful Transition: An International Delphi Study." *Journal of Adolescent Health* 56 (6): 612–18.

frameworks for adolescent care are Jamaica and the United States. Health care systems in the United States have a distinct separation between pediatric care and adult care facilities, particularly in SCD patients. When patients move towards adult care, they often encounter myriad psychosocial issues regarding disease knowledge, self-care, coping, readiness for independence, and the movement from a pediatric center to an adult center.[85] Due to the nature of this system there is a formalized structure for the transition to adult care. In Jamaica however, the concept of transition does not exist, as SCD patients receive clinical care using a comprehensive model. The same psychosocial issues as well as other disease complications that patients may face are all addressed by pediatricians, general family physicians, and hematologists in the same center.[86]

Readiness, or transition readiness, can be described as specific decisions made and actions taken in building the capacity of the adolescent, family, and providers, to begin, continue and finish the process of transition. In both Jamaica and the United States, disease knowledge and readiness are essential for adolescents to be able to

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[85] Telfair, Joseph, Leah R. Alexander, Penny S. Loosier, Patty L. Alleman-Velez, and Julie Simmons. 2004. "Providers' Perspectives and Beliefs Regarding Transition to Adult Care for Adolescents with Sickle Cell Disease." *Journal of Health Care for the Poor and Underserved* 15 (3): 443–61.

[86] Bhatt, Komal, Marvin E. Reid, Norma A. Lewis, and Monika R. Asnani. 2011. "Knowledge and Health Beliefs of Jamaican Adolescents with Sickle Cell Disease." *Pediatric Blood & Cancer* 57 (6): 1044–48.

independently manage their illness apart from their parents or guardians in order to reduce long term complications.[87, 88]

1.6 Study Aims

The research goal for this project is to compare the two systems that exist for treatment of adolescents with SCD in Jamaica and the United States by assessing differences and similarities in patients' readiness for adult treatment and their understanding of SCD and its management. The 4 main aims of this study are: (1) to identify individual disease knowledge and readiness for adult care, (2) to investigate the similarities and differences in adolescent patient disease knowledge and readiness for adult care as a result of contrasting frameworks in Jamaica and the United States, (3) to understand patient attitudes towards beginning and receiving adult care, as well as the issues and barriers they may encounter, and (4) to investigate the applicability of standardized methods of transition assessment that are currently used in the United States in an international setting.

Understanding when and how involved a patient will be as they begin to move into adult care is one of the most difficult aspects of transition. There are several

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[87] Telfair, Joseph, Leah R. Alexander, Penny S. Loosier, Patty L. Alleman-Velez, and Julie Simmons. 2004. "Providers' Perspectives and Beliefs Regarding Transition to Adult Care for Adolescents with Sickle Cell Disease." *Journal of Health Care for the Poor and Underserved* 15 (3): 443–61.

[88] Bhatt, Komal, Marvin E. Reid, Norma A. Lewis, and Monika R. Asnani. 2011. "Knowledge and Health Beliefs of Jamaican Adolescents with Sickle Cell Disease." *Pediatric Blood & Cancer* 57 (6): 1044–48.

methods to assess readiness. For purposes of this study, the American Society of Hematology's (ASH) *Sickle Cell Disease Transition Readiness Assessment,* released in 2016 was used alongside an age-stratified disease knowledge questionnaire. Questionnaires were administered and in-depth interviews conducted with adolescents with sickle cell disease in Jamaica, and results from the questionnaires were analyzed alone and compared to the same questionnaires that have been done in the United States as part of standard practice. The goal was to reach a target of 50 completed surveys, and age match them to previously completed patient surveys at Duke University. The interviews were done in Jamaica to offer a better understanding of the beliefs and perspectives of adolescent patients, an in-depth look at individual disease knowledge, and give more information on the strengths and weaknesses that are unique and similar in Jamaica and the United States. Defining patient perspectives exposes the barriers and challenges that patients may face and increases comprehension of the existing relationship between adolescents and health care. The target number for in-depth interviews in Jamaica was between 10-20 patients.

Results from this study will provide insight for further action to address urgent needs and improve existing practices in the future. Furthermore, the information gathered throughout this study may have implications on program development, public health, and future research.

2. Methods

2.1 Settings

Jamaica is an island located in the Caribbean with a population of 2.8 million people. The official language is English; however, natives speak in their own dialect called Patois. This island is also host to many cultural and religious beliefs, including several denominations of Christianity, Hinduism, Judaism, Islam, and Rastafarianism. A majority of the population, 48%, is under 30 years old and the gross national income (GNI) per capita in 2016 was 4630 USD.[1] SCD remains prevalent in the region. Screening of 100,000 consecutive non-operative deliveries in the Jamaica Cohort Study found sickle cell trait in 10.0% and HbC trait in 3.6%. Incidence of HbSS was found in 1/300 births, HbSC in 1/500 births, $HbSB^+$ in 1/3000 births, and $HbSB^0$ in 1/7000 births.[2] This study was conducted in the Sickle Cell Unit at the University of West Indies (UWI) in Kingston, Jamaica.

In the United States, particularly in North Carolina, there were approximately 5,578 individuals with SCD between 2004 and 2008, and 92 newborns were born with the condition, 27 of them in Durham County. In North Carolina, SCD occurs in 1 out of every 1,435 births, and 1 out of every 360 African American births. However, SCD

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[1] The World Bank. 2016. "Jamaica Country Profile." https://data.worldbank.org/country/Jamaica

[2] Serjeant, Graham R. 2013. "The Natural History of Sickle Cell Disease." *Cold Spring Harbor Perspectives in Medicine* 3 (10). https://doi.org/10.1101/cshperspect.a011783.

occurs in all racial and ethnic groups in North Carolina including Hispanics and American Indians.[3] The comparison of previously completed, anonymized questionnaires and surveys with those done in Jamaica occurred at the Sickle Cell Center in Duke University Hospital.

2.1.1 Participants

Eligibility for this study was defined as patients with SCD, either male or female, seeking treatment at the health facility in the University of the West Indies, having no acute illness at the time of study, and were between the ages of 13-19. Additionally, if the patient was below the local legal age of consent of 16, both the parent and the participant must have completed both written and verbal informed consent process. Recruitment for this study occurred in the Sickle Cell Unit at the time of check-in, and the consent and data collection procedures were done during the wait period before the doctor's visit.

Duke University patients questionnaires and surveys follow most of the same eligibility criteria as Jamaican participants: patients with SCD, either male or female, between the ages of 13-19. However, the surveys and questionnaires that were used

[3] The Center for Disease Control. Sickle Cell Disease in North Carolina. 2016. "CDC Registry and Surveillance System for Hemoglobinopathies (RuSH)." https://www.cdc.gov/ncbddd/sicklecell/documents/scd_in_nc_providers.pdf)

must have been completed at the Duke University Sickle Cell Center, as all information was collected anonymously and after completion.

2.2 Procedure

In Jamaica, recruitment and participation in the study occurred during the same doctor's visit. Eligibility to participate was determined by nursing staff during the standard check-in procedures. While checking vitals and assessing physical fitness, nurses screened adolescents within the appropriate age range and, if willing to discuss the study, patients were referred to the researcher. The consent process was done in the same private room as data collection, and all participants were given the option to take part in either the questionnaire portion, or both the questionnaire and interview portions of the study. Once consent forms were read and signed, the parents/guardians, if present, were asked to leave. Participants first completed a demographic survey. All questions on the demographic survey were asked verbally and responses were simultaneously written by the researcher into the corresponding spaces. After demographic information was collected, the participants were then asked to complete the Transition Readiness Assessment and the age-appropriate disease knowledge questionnaire. If participants elected to take part in the interviews during the consent process, interviews were done immediately after the questionnaires and recorded for transcription. There was no follow-up procedure.

Duke University Sickle Cell Center patient questionnaires and surveys were previously completed prior to the start of this study and were done as part of standard practice. As a result, aside from age, no demographic information was collected, and these surveys were anonymous at the time of data collection. The surveys were randomly sampled and age-matched, within the age range of the study of 13-19.

All study procedures were approved by the institutional review boards at Duke University and University of the West Indies.

2.3 Measures

In Jamaica, each participant completed a demographic survey, disease knowledge questionnaire, and the ASH Transition Readiness Assessment Questionnaire. This was the first effort to use both the Transition Readiness Assessment and disease knowledge questionnaires in this setting. The demographic survey served as a tool to collect information about the study population. Data collected on the demographic survey included age, gender, race/ethnicity, employment, education level, religion, insurance status, existence of relatives with SCD, and marital status of parents. Questions relating to socioeconomic information asked about the number relatives living at home, with a subsection specifying who lived at home with the participant, number of rooms at home, and the possession of a list of items such as air conditioning, fan, telephone, television, computer, internet, etc.

Developed by Dr. Shah at Duke University as a standard of care assessment, the disease knowledge questionnaires used in this study were age-stratified into three groups, ages 13-14, 15-16, and 17-19. Each questionnaire was comprised of 10 questions, increasing in difficulty with age, that gauged the level of disease knowledge. Questions assessed age appropriate disease knowledge based on teaching and included question such as: which kind of SCD the participant had, what "sickling" meant, what hydroxyurea was, and how to manage the disease. The questionnaires for 13-14 year-olds focused more on disease knowledge, while management was introduced in 15-16 year olds, and expanded in 17-19 year olds. Specification on the SCD genotype were only on the questionnaires corresponding to the youngest and oldest groups. Questions pertaining to health insurance were only asked of the 17-19 age group.

ASH released an assessment in 2016 as a tool to ease the transition into adult care by identifying and eliminating gaps in care, starting the conversation between parents, patients, and providers, and help patients individually manage health care .[4] The assessment had 5 sections: disease knowledge, medication management, appointments, and private insurance, with answers confirming or denying activities pertaining to each category. Answers were measured quantitatively through the responses "no, I do not

[4] "ASH Releases Toolkit for Transition from Pediatric to Adult Hematologic Care." 2017. September 7, 2017. /Advocacy/Policy-News/2016/5581.aspx.

know," "no, but I am learning to do this," "yes, I have started doing this," and "yes, I always do this when I need to."

Interviews questions were a combination of original questions that expanded on the questionnaires, as well as taken from previous sources (Williams et al, 2011)[5], and fit to the study location and purpose. The questions were divided into 3 sections: disease knowledge and medication management, involvement with treatment and disease management, and personal feelings and experiences with the health care environment. The audio-recorded interviews served to reinforce and expand on questionnaires, as well as provide an in-depth understanding of their disease knowledge and their readiness to receive adult care.

2.4 Analysis

This is mixed-methods study that incorporates both qualitative and quantitative analysis procedures. Mean values were generated for disease knowledge questionnaires and stratified by age, gender and socioeconomic status (SES) factors. A p-value for differences in test scores between age groups, gender and SES was estimated using Fisher's exact test. Transition Readiness Assessments sections were analyzed independently. A p-value for difference in assessment responses between age groups

[5] Williams TS, et al. 2011. "Measurement of medical self management and transition readiness among Canadian adolescents with special health care needs." *International Journal of Child and Adolescent Health 527-35*

was estimated using a Kruskal-Wallis test. Applicability for the ASH Transition Readiness Assessment was defined as concordance with corresponding interview responses in relation to comparison with data from Duke University. Interview responses were analyzed by section using content analysis, identifying common themes overall, and among age groups. Important or interesting findings or text were extracted for purposes of data dissemination.

3. Results

The sickle cell clinic at UWI serves patients from infancy into late adulthood, and of the people who fit eligibility criteria, exactly 50 were successfully recruited to take part in the study. A total of 20 participants also elected to take part in the recorded in-depth interviews after the consent process, however 2 chose to end early, resulting in 18 complete interviews.

3.1 Study Population

Table 1 shows demographic information that was collected via the demographic survey. The age groups were represented evenly, while females made up 58% of all participants. The majority self-identified as black, however 24% chose to specify that they were of mixed race. The overwhelming majority of all participants had completed up to a secondary level education at the time of data collection. Only 5 people were employed, on a part-time basis. None indicated that they had, at any point, worked full-time. A total of 56% of participants were enrolled in private health insurance, while the remainder paid out of pocket for health services. The demographic survey also asked if any family members had SCD, and while many knew of relatives who had the trait, only 21 participants could identify a family member with SCD in either their nuclear or extended family. Concerning religion, most participants, 86%, were affiliated with Christianity, and one person identified as Christian on her mother's side, and Muslim on her father's side. Only 28% of participants had parents who were together.

Table 1: Demographic Information for All Participants in Jamaica (N=50)

Participants		n (%)
Age		
	13-14	16 (32)
	15-16	16 (32)
	17-19	18 (36)
Gender		
	Male	21 (42)
	Female	29 (58)
Race/Ethnicity		
	Black	38 (76)
	Other (Mixed)	12 (24)
Education		
	Primary	3 (6)
	Secondary	40 (80)
	Post-Secondary	4 (8)
	University	3 (6)
Employment (age 16+, n = 26)		
	Unemployed	21 (80.77)
	Part-Time	5 (19.23)
	Employed	0
Religion		
	Christian	43 (86)
	Christian/Muslim	1 (2)
	None	6 (12)
Payment		
	Out of Pocket	22 (44)
	Private Health Insurance	28 (56)
Family with SCD		
	Nuclear	9 (18)
	Extended	11 (22)

	Nuclear & Extended	1 (2)
	None	29 (58)
Marital Status of Parents		
	Together	14 (28)
	Separate	34 (68)
	One/Both Deceased	2 (4)

Table 2 indicates responses to specific questions regarding socioeconomic information on the demographic survey. When asked about family living at home, participants were asked to identify who lived with them at that time, including mother, father, stepmother, stepfather, siblings, grandparents, aunts, uncles, cousins, and any other relatives or non-relatives. Of all participants, 86% lived with their mothers, and 32% lived with their fathers. From those who lived with a parent, 13 participants reported living with *both* their mother and father, while 4 responded not living with either parent. Number of items at home was a count of a list of items on the demographic survey where participants responded either yes or no to possession. Items included sewing machine, stove, refrigerator, air conditioner, fan, microwave, telephone, radio, stereo, CD player, VCR/DVD, washing machine, television, cable, satellite, bicycle, motorcycle, car, computer, and internet. Responses are reported quantitatively in categories. In total, 52% reported having 11 to 15 items on the list; 76% had internet, 82%

had a computer, and 52% had a car or some means of transportation (other than a motorcycle, which corresponded to only 8% of participants).

Table 2: Socioeconomic Information for All Participants in Jamaica (N=50)

Participants		n (%)
Family living at home		
(not including respondent)	1 to 3	22 (44)
	4 to 6	16 (32)
	7 to 9	7 (14)
	10 to 12	5 (10)
Number of rooms		
	1 to 6	34
	7 to 12	16
Number of items owned		
	6 to 10	12 (24)
	11 to 15	26 (52)
	16 to 20	12 (24)

Table 3 expands on the level of crowding that participants experienced at home. A majority of participants, 22 people, reported only having 1 to 3 family members living with them, and well over half of them had between 1 and 6 rooms at home. Only 3 people reported evidence of extreme crowding, i.e. reporting 10 to 12 people living at home and 1 to 6 rooms.

Table 3: Number of Rooms Stratified by Family Members Living at Home

Rooms	Number of Family Members at Home (%)	Total

	1 to 3	*4 to 6*	*7 to 9*	*10 to 12*	
1 to 6	15 (44.12)	10 (29.41)	6 (17.65)	3 (8.82)	34
7 to 12	7 (43.75)	6 (37.5)	1 (6.25)	2 (12.5)	16
Total (%)	22 (44)	16 (32)	7 (14)	5 (10)	50

3.2 Quantitative Findings

3.2.1 Disease Knowledge Questionnaire

Age, gender, and socioeconomic factors are considerations that influence disease knowledge readiness for transition.[1] A Fisher's exact test was used to estimate a p-value for differences in each of these variables, at an alpha level 0.05.

Tables 4 and 5 report mean scores by age group and by gender, as well as frequency for each score. The overall average score for all disease knowledge questionnaires was 5.9. No participant answered all 10 questions correctly, or scored below a 3. Those ages 15-16 had a higher mean score, 6.5, as well as the most people score a 7 or above. Conversely, age group 17-19 had the lowest mean scores, 5.44, and no participant answered 9 questions correctly. A majority scored either 5 or a 6 in all 3 age groups, but the highest number of participants who received scores in this range was in age group 17-19. More people in age group 13-14 scored a 3 than in the older age groups. Average scores between males and females were very similar, 5.95 and 5.86 respectively and scores were distributed relatively evenly across the test range.

Table 4: Disease Knowledge Questionnaire Total Scores Stratified by Age

Age Group	Mean	Frequency Score (%)							Total
		3	4	5	6	7	8	9	
13-14	5.81	4 (25)	1 (6.25)	0	6 (37.5)	1 (6.25)	2 (12.5)	2 (12.5)	16
15-16	6.5	2 (12.5)	1 (6.25)	3 (18.75)	2 (12.5)	2 (12.5)	1 (6.25)	5 (31.25)	16
17-19	5.44	1 (5.56)	3 (16.67)	6 (33.33)	5 (27.78)	1 (5.56)	2 (11.11)	0	18
Total	*5.9*	7 (14)	5 (10)	9 (18)	13 (26)	8 (4)	5 (10)	7 (14)	50
Fisher's Exact Test					*p = 0.079*				

Table 5: Disease Knowledge Questionnaire Total Scores Stratified by Gender

Gender	Mean	Frequency Score (%)							Total
		3	4	5	6	7	8	9	
Male	5.95	3 (14.29)	2 (9.52)	3 (14.29)	6 (28.57)	2 (9.52)	2 (9.52)	3 (14.29)	21
Female	5.86	4 (13.79)	3 (10.34)	6 (20.69)	7 (24.14)	2 (6.9)	3 (10.34)	4 (13.79)	29
Total	5.9	7 (14)	5 (10)	9 (18)	13 (26)	4 (8)	5 (10)	7 (14)	50
Fisher's Exact Test					*p = 0.998*				

All socioeconomic factors present on the demographic survey and the corresponding mean knowledge scores of the participants are reported in Table 6. No significant associations were shown relative to any of the information; however, more participants scored higher, a mean value of 6.5, when they had a moderate number of family living at home, 4 to 6 people, as opposed to 10 to 12 people. Additionally, possessing more items as home was correlated with a higher mean score, an average of 6.42 among 12 people who owned between 16 and 20 items. The average score for those with one or both deceased parents is 7; however, this applies to only two participants.

Having more rooms at home (7-12) related more to a lower mean score than having fewer rooms (1-6), 5.57 and 6.06, respectively.

Table 6: Disease Knowledge Questionnaire Mean Scores Stratified by Socioeconomic Factors and Parents' Marital Status

Variable		Mean	Fisher's Exact Test
Rooms			***p = 0.276***
	1 to 6	6.06	
	7 to 12	5.57	
Items at Home			***p = 0.694***
	6 to 10	6	
	11 to 15	5.62	
	16 to 20	6.42	
Family living at Home			***p = 0.182***
	1 to 3	5.82	
	4 to 6	6.5	
	7 to 9	5.42	
	10 to 12	5	
Parents			***p = 0.17***
	Together	5.71	
	Separate	5.91	
	One/Both Deceased	7	

A question asking about sickle cell type was present on the questionnaire corresponding to age groups 13-14 and 17-19. On the 13-14 questionnaire, the question was phrased, "what type of sickle cell disease do you have?" while ages 17-19 were

asked, "what is your hemoglobin type?". Answers were cross referenced with information about each patient's sickle cell type that is available on their medical record, and is reported in Table 7 as a right or wrong response. The Fisher's exact test estimated a p-value of 0.681, which suggests that there is no statistically significant difference in the number of incorrect responses between both age groups, however, more participants ages 17-19, 83% as opposed to 75%, who responded correctly.

Table 7: Disease Knowledge Questionnaire Responses on Sickle Cell/Hemoglobin Type Between Patients Ages 13-14 and 17-19[1]

Age Group	Wrong Responses (%)	Right Responses (%)	Total
13-14	4 (25)	12 (75)	16
17-19	3 (16.67)	15 (83.33)	18
Total	7	27	34
Fisher's exact test		***0.681***	

3.2.1.1 Comparison with Duke University Sickle Cell Center

Disease knowledge questionnaires by participants from Jamaica ages 17-19 were compared to composite scores by patients from Duke University Sickle Cell Center. As a whole, no patients from Duke scored below an 8, and 4 patients were able to receive a perfect score of 10. The mean was an 8.75, which is a 62% increase from the mean score of participants in Jamaica in the same age group.

[1] Question about sickle cell type was only asked in the Disease Knowledge Questionnaires corresponding to the youngest (13-14) and oldest (17-19) age groups

Table 8: Comparison of Disease Knowledge Total Scores from Duke Patients with UWI Patients

	Mean	SD	Frequency Score (%)								Total
			3	4	5	6	7	8	9	10	
UWI SCU	5.44	1.34	1 (5.6)	3 (16.7)	6 (33.3)	5 (27.8)	1 (5.6)	2 (11.1)	0	0	18
DU SCC	8.75	0.86	0	0	0	0	0	8 (50)	4 (25)	4 (25)	16
% Difference of Means				***62.17***							

3.2.2 ASH Transition Readiness Assessment

All Transition Readiness Assessment responses were given numerical values between 0 and 4, with no/missing response as 0, "no, I do not know" as 1, "no, but I am learning to do this" as 2, "yes, I have started doing this" as 3, and "yes I always do this when I need to" as 4. Responses were added together in a composite score for each section on the questionnaire, and stratified by age. A Kruskal -Wallis test estimated a p-value a difference between all age groups, and youngest and oldest participants, at an alpha level of 0.05.

The disease knowledge section had 9 questions in total, with a possible minimum score of 9, and maximum score of 36. As shown in Table 9, and again visually in Figure 1, all participants' confidence in disease knowledge increased with age, even reaching the highest possible score in the 17-19 age group. Both minimum and maximum scores increased, and mean scores steadily increased from 22.13 at age 13-14

to 26.27 at age 17-19, representing significantly different 18.71% increase between the youngest and oldest participants, with an estimated p-value of 0.02.

Table 9: Transition Readiness Assessment Stratified by Age (Jamaica): Disease Knowledge

Age	Mean	SD	Min	Median	Max	Q1	Q3
13 - 14	22.13	4.54	13	24	27	19	25.5
15 - 16	23.81	4.5	16	24	33	18.5	28
17 - 19	26.27	4.7	18	26	36	24	29
Kruskal-Wallis Test							
Total			***p = 0.073***				
Youngest vs. Oldest			***p = 0.023***				

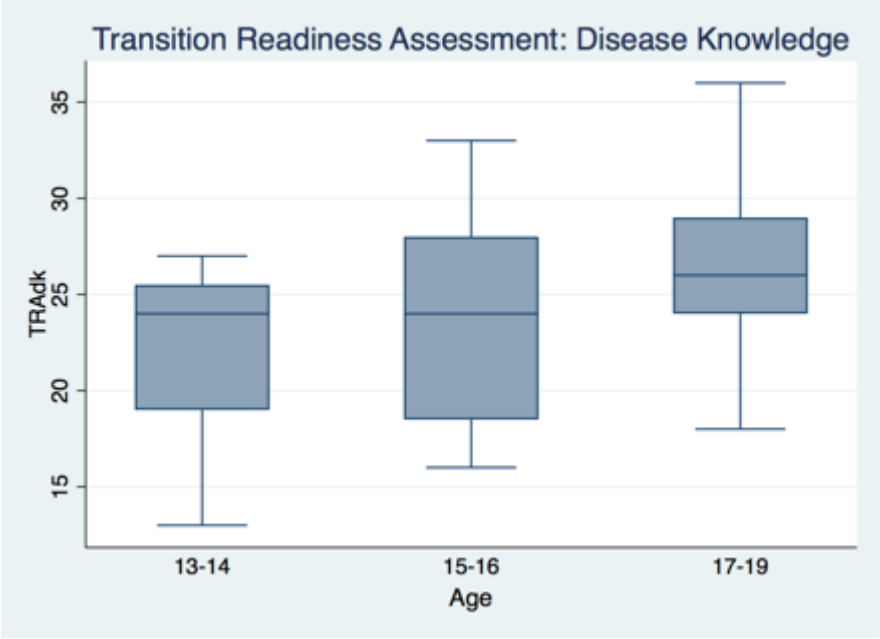

Figure 1: Transition Readiness Assessment Sums by Age Regarding Questions About Disease Knowledge

Table 10 shows the composite scores for questions about medication management. The section involves 6 questions; the highest possible score was 24 and the lowest was 6. Mean scores rose slightly with age, as did maximum scores. Minimum scores overlapped slightly between all three groups, with a drop in minimum scores in the middle age group, 15-16 years. Differences in scores were not significant.

Table 10: Transition Readiness Assessment Stratified by Age (Jamaica): Medication Management

Age	Mean	SD	Min	Median	Max	Q1	Q3
13 - 14	17.63	2	14	17	21	16	19
15 - 16	17.94	2.62	13	18	23	16.5	19.5
17 - 19	18.5	2.95	14	19	24	16	20
Kruskal-Wallis Test							
Total			***p = 0.676***				
Youngest vs. Oldest			***p = 0.427***				

The appointments section is the largest and most significant improvement with age, and findings are presented in Table 11. There were 8 questions on the assessment about appointments. The maximum possible score was 36, while the minimum score was 8. The self-evaluations for this section increased significantly with age, with the mean increasing from 15 to 20.22, with the overall estimated p-value of 0.012, and 0.006 when comparing scores for the youngest and oldest age groups. Interestingly, minimum scores were similar for 13-14 and 17-19, at 8 and 9 respectively. Maximum scores jumped from 19 to 29, although no one reached 36. Figure 2 shows the overall jump in

confidence for booking, getting to, and reaching out for appointments and medical health care, as well as the overlap in composite scores.

Table 11: Transition Readiness Assessment Stratified by Age (Jamaica): Appointments

Age	Mean	SD	Min	Median	Max	Q1	Q3
13 - 14	15	3.22	8	15.5	19	13.5	17.5
15 - 16	16.63	3.63	11	17.5	24	14	19
17 - 19	20.22	5.79	9	20	29	15	26
Kruskal-Wallis Test							
	Total		***p = 0.012***				
Youngest vs. Oldest			***p = 0.006***				

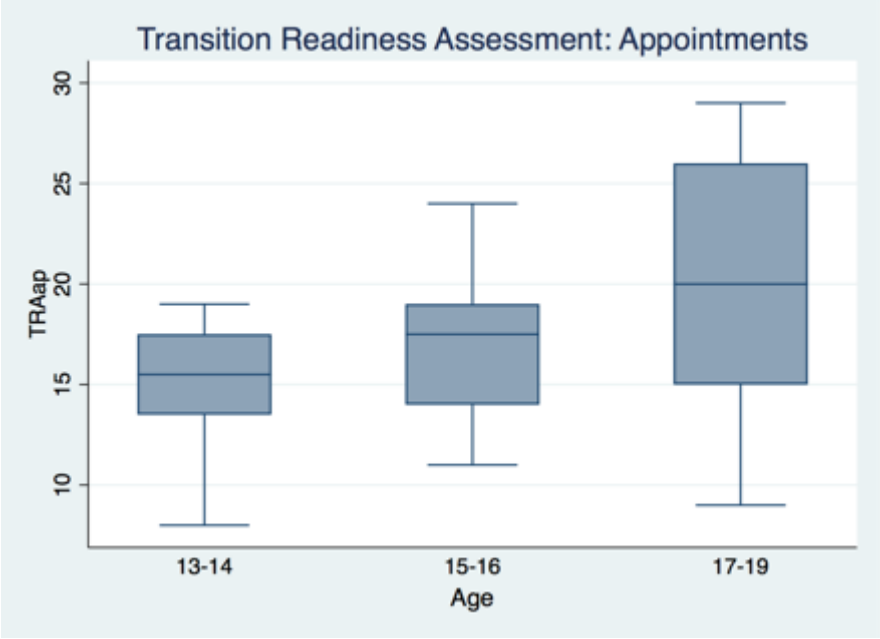

Figure 2: Transition Readiness Assessment Sums by Age Regarding Questions About Appointments

Table 12 reports the composite scores for the 2 questions that make up the insurance section of the assessment, "I carry my own insurance card" and "I understand my insurance plan." There was no significant difference in the assessment between age groups. All three age groups had minimum scores at 2, which can be attributed to the variation in participants with private health insurance and those who paid out-of-pocket. Maximum scores increased with age, from 6 to the maximum possible score of 8, indicating a clearer understanding for how insurance functions. There was a drop in mean for age group 15-16 from 3.13 at age 13-14 to 2.56, and the maximum score was 4, also lower than the younger age group, where the minimum was 2.

Table 12: Transition Readiness Assessment Stratified by Age: Insurance

Age	Mean	SD	Min	Median	Max	Q1	Q3
13 - 14	3.13	1.5	2	2	6	2	4
15 - 16	2.56	0.73	2	2	4	2	3
17 - 19	3.61	2	2	3	8	2	5
Kruskal-Wallis Test							
Total			***p = 0.384***				
Youngest vs. Oldest			***p = 0.479***				

The privacy information section was only one question, "I understand how health care privacy changes at 18, when I am legally an adult". The results in Table 13 shows there was an increase in understanding with increasing age, with and estimated p-value of 0.053 between 13-14 and 17-19 years old. The maximum value for the

youngest participants was 3, indicating some had started to learn, while the 17-19 age group still had individuals who did not understand changes in their health care privacy, although they were considered adults in Jamaica. This may be due to differences in health care systems.

Table 13: Transition Readiness Assessment Stratified by Age: Privacy Information

Age	Mean	SD	Min	Median	Max	Q1	Q3
13 - 14	1.81	0.83	1	2	3	1	2.5
15 - 16	2.13	1.09	1	2	4	1	3
17 - 19	2.56	1.1	1	3	4	2	3
Kruskal-Wallis Test							
Total			***p = 0.156***				
Youngest vs. Oldest			***p = 0.053***				

3.2.2.1 Comparison with Duke University Sickle Cell Center

Composite scores for participants from Duke University were compared to the results from participants in Jamaica, and compiled in Table 14 as well as presented visually in Figure 3. There were 13, 18, and 19 patients in age groups 13-14, 15-16 and 17-19, respectively. Differences are signified by a percent (%) change, either positive or negative relative to Jamaica, in the mean for each section of the assessment. All sections, for each age group show Duke patients have increased overall scores in self-assessment, apart from *appointments* in age group 13-14 (-12.11%), and *insurance* in age group 13-14 (-26.2%). The largest differences are in the older age groups where transition to adult care

usually commences or occurs, in *disease knowledge* in age groups 13-14 (26.26%) and 15-16 (29.02%), *privacy information* in age group 15-16 (38.03%) and *insurance* in age group 17-19 (37.12%).

Table 14: Comparison of ASH Transition Readiness Assessment Results from Duke Patients and Jamaica Patients

Age		Duke SCC	UWI SCU	% Diff of Means		Duke SCC	UWI SCU	% Diff of Means
	Disease Knowledge				Medication Management			
13 - 14								
	Mean	27.92	22.13	26.16	*Mean*	20.31	17.63	15.2
	Min	16	13		*Min*	12	14	
	Median	30	24		*Median*	21	17	
	Max	36	27		*Max*	24	21	
	Q1	25	19		*Q1*	20	16	
	Q3	36	25.5		*Q3*	23	19	
15 - 16								
	Mean	30.72	23.81	29.02	*Mean*	19.89	17.94	10.87
	Min	21	16		*Min*	13	13	
	Median	30.5	24		*Median*	21	18	
	Max	36	33		*Max*	24	23	
	Q1	28	18.5		*Q1*	18	16.5	
	Q3	35	28		*Q3*	24	19.5	
17 - 19								
	Mean	30.58	26.27	16.1	*Mean*	20.68	18.5	11.78
	Min	20	18		*Min*	8	14	
	Median	32	26		*Median*	21	19	
	Max	36	36		*Max*	24	24	
	Q1	27	24		*Q1*	19	16	

	Q3	36	29		*Q3*	24	20	
	Appointments				**Privacy Information**			
13 - 14								
	Mean	13.38	15	-12.11	*Mean*	1.92	1.81	6.08
	Min	8	8		*Min*	1	1	
	Median	13	15.5		*Median*	2	2	
	Max	19	19		*Max*	4	3	
	Q1	10	13.5		*Q1*	1	1	
	Q3	17	17.5		*Q3*	2	2.5	
15 - 16								
	Mean	17.72	16.63	6.15	*Mean*	2.94	2.13	38.03
	Min	10	11		*Min*	1	1	
	Median	17.5	17.5		*Median*	3.5	2	
	Max	30	24		*Max*	4	4	
	Q1	14	14		*Q1*	2	1	
	Q3	21	19		*Q3*	4	3	
17 - 19								
	Mean	22.21	20.22	9.84	*Mean*	2.74	2.56	7.03
	Min	11	9		*Min*	1	1	
	Median	21	20		*Median*	3	3	
	Max	32	29		*Max*	4	4	
	Q1	20	15		*Q1*	2	2	
	Q3	25	26		*Q3*	4	3	
	Insurance							
13 - 14								
	Mean	2.31	3.13	-26.2				
	Min	2	2					
	Median	2	2					
	Max	4	6					
	Q1	2	2					

	Q3	2	4	
15 - 16				
	Mean	2.89	2.56	12.89
	Min	2	2	
	Median	3	2	
	Max	5	4	
	Q1	2	2	
	Q3	3	3	
17 - 19				
	Mean	4.95	3.61	37.12
	Min	2	2	
	Median	5	3	
	Max	8	8	
	Q1	2	2	
	Q3	8	5	

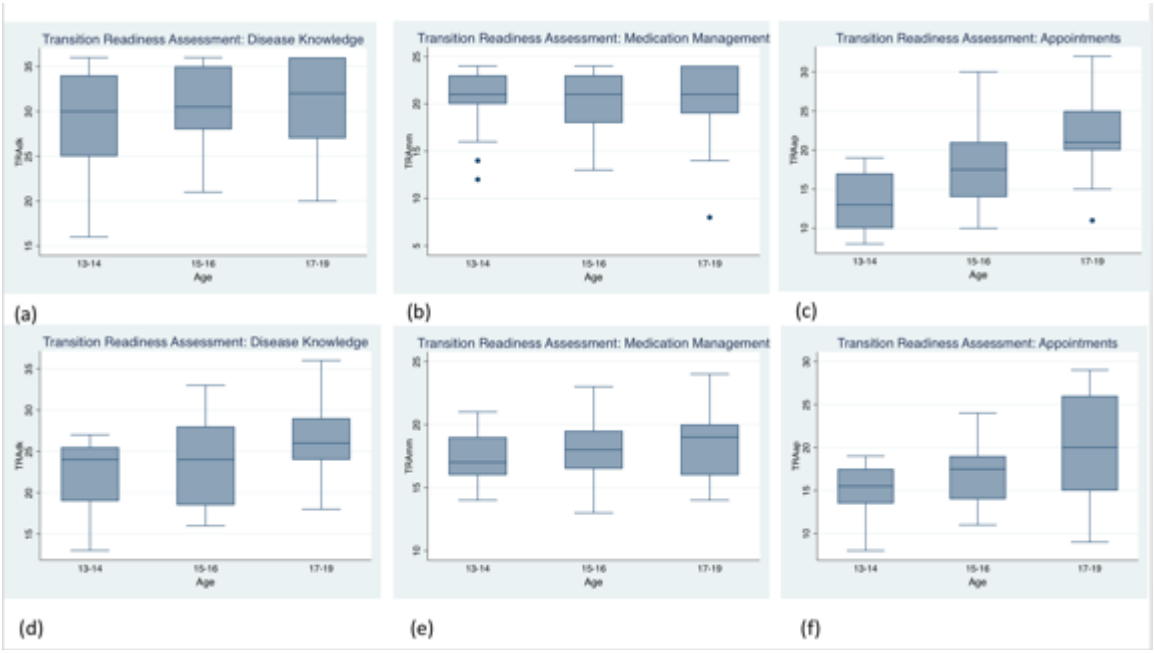

Figure 3: Box Plots Comparing Transition Readiness Assessment Scores for Disease Knowledge, Medication Management, and Appointments for Patients at DU SCC (a-c) and UWI SCU (d-f)

3.3 Qualitative Findings

In-depth interviews were conducted with participants from different age groups. The questions asked about disease knowledge, medication and medication management, involvement in disease management, and personal patient experiences. Participants were free to answer as they saw fit, and responses varied in knowledge and self-reliance.

3.3.1 Disease Knowledge, and Medication Management

The interview began with a question to understand what each respondent knew about SCD, which could be general knowledge, or anything that applied personally to their disease experience. They were subsequently asked about their specific health care

needs. When asked about sickle cell disease, many responded with their personal experiences first. Many also brought up how cold weather and dehydration were a main aspect of their disease knowledge, claiming always to stay warm, drink water, exercise, and eat healthy.

> "Well, I know sickle cell disease is really dangerous but some ways to avoid getting sick and pain because you know it accompany with a lot of belly aches and so on and […] when the climate's cold you keep warm, you're wrapped up and you put on, take jackets, keep warm because like when climate is still cold it breaks the cell up and so on and what I normally […] always drink some bitter bush helps a lot like when my eye get kind of yellow it kind of clear it and so on." – *female, 15-16*

One respondent described an experience with cold weather, while simultaneously identifying her hemoglobin type, then went on to explain how it differentiated her experiences with what she knew about SCD.

> "Well I am HBSS and me really, I don't really get sick a lot but what triggers me getting sick is climate change. Like for example it's hot […] then all of a sudden I get cold, I get fever and I have to go to the doctor and the last time I went to the doctor was because my bone marrow had stopped making red blood cells because I had some type of infection so I had to stay in the hospital and got blood, like three packs for blood transfusion. I was shocked. I was scared." – *female, 17-19*

Several participants described the basic disease pathology immediately after being asked about their knowledge of SCD. However, knowledge of inheritance patterns was not consistent among everyone interviewed.

> "Okay so I know that I have the SC type sickle cell disease which is brought down from the recessive normal gene and the dominant sickle gene. I'm not really sure from which parent actually. I know that sickle cell disease is hereditary as you know I guess before, it's a disease of the red blood cells which cause them to be sickle shaped and block arteries in the body causing pain because of a lack of oxygen in that area of the body. I know that pain isn't the

> only complication that is associated with sickle cell. I know that acute chest syndrome or popping or squeaking or any other illness or complication that might have to do with the bone[…].” – *male, 15-16*

There were also several participants who did not have a solid grasp on disease knowledge, and had difficulty articulating answers about what they felt they knew. One individual knew that the disease was inherited, and knew about phenotypic variation, but phrased it through concerns about future children inheriting the condition.

> “The actual disease, not much, but I know that you can't get a girlfriend with the same disease because that would cause problems later on, you know like if you want to have a baby or whatever. […] Because the child would be born with it and you don't know how they would react because not everyone is the same.” – *male, 17-19*

Participants who belonged to the 13-14 age groups were generally dependent on their parents for the entirety of their disease knowledge.

> “Well do not know anything because my daddy keep track of it and my mommy, they don't tell me anything. […] I just know like if you're feeling pain he'll go for like the Cataflam or the Panadol or anyone of those stuff. I seen the bottle I take.” – *female, 13-14*

Knowledge about management of health care and medicine increased with age. Those who were older could provide specific scenarios to describe their health care needs, and even cited which medicines to take with each situation, and the doses that accompanied them. One participant even cited the problems that are associated with opiate pain relievers.

> “So, so far I've gotten like Cataflam like fifty milligrams. First I started at twenty five, no before I started on liquid and on to 25 milligrams and then 50 milligrams now and then they give you a lot of things like things like the other day when I was sick, they gave me like Voltaren, I don't even remember the dosage but it

> was high and they give you like morphine when it's really bad but I'm not a big fan of those because it really can make you develop problems later on in your life so I'm not a big fan of them, but I mostly take Cataflam." – *male, 17-19*

Younger participants usually cited the involvement of their parents when describing their needs in managing their SCD.

> "Well I take a like, I mean when I'm in sickle cell pain or once I start feeling, if I'm at school and I'm feeling pain, find the nurse and then nurse treat and give me medication like Panadol and I will stay there, call my parents and they come get me and bring me to the nearest hospital. When the […] treat me, watch me till I feel better, […] then they send me home, give me a letter to come to sickle cell unit and they check me up if I have any blood test to do I do that, if I have an injection to get, I get it." – *female, 13-14*

However, one participant felt they had no frame of reference for their disease and could not answer the question with a verbal response. What constituted a "need" did not just include medicines, but also lifestyle behaviors like personal effort and responsibility, social interaction, and exercise.

> "Well my healthcare needs is basically me trying my best to take care of myself so as I said before like the exercises and I try to be social as well so my healthcare needs so I have people that I can talk to but it's not something that you really bring up in a conversation so that's one and then you taking tablets so that you stay on top and basically just taking care of yourself. That's really it." – *male, 17-19*

Disease knowledge questions also asked participants to explain what they knew about SCD and puberty. Most did not know how SCD affected development and puberty, particularly the youngest participants, apart from slowing the process, referring to growth and size as the main indicator for an adolescent with SCD.

> "Well I know that it's a disease where it affects the growth in children you know like children will grow like twice my size." – *male, 13-14*

Male participants were more aware of how SCD affected them. Two participants, aged 18 mentioned that lack of facial hair was one of the main experiences they noticed.

> "[…] there is a lot of things that should come with puberty that haven't really come with puberty so I'm aware that it has slow down my growth and my facial hair, that's one, but it's coming!" – *male, 17-19*

Several participants could not explain how drugs, tobacco, and alcohol affect SCD, but all agreed that refraining from consuming them was best for their condition, some even saying that they did not know the answer because they never experimented with them.

> "I don't take drugs or alcohol. […] You know I really don't know how it affect I just know that I came under like you're not supposed to take it. I know smoking can give you lung cancer […]." – *female, 17-19*

Only one participant explained specifically explained how alcohol can contribute to complications with SCD.

> "[…] drinking though, the drinking dehydrates you so if you don't drink enough water while drinking it can cause a pain series on you there." – *male, 17-19*

Participants were then asked if they felt they had the right information about their health and their health care. Responses varied in the level of initiative and personal responsibility. Many showed the interest in wanting to learn, as well as addressed the limitations in their knowledge and how they may go around finding solutions to filling those gaps.

> "Not really. I feel like I should get more information about it. […] Like I think I myself should like, cause there's internet search more about it, the effect and a lot of things about it." – *female, 15-16*

> "I mean I feel so, but sometimes you don't know because you don't know everything and then when something else happens you don't know what could cause that so I guess that's why you have doctors there." – *male, 17-19*

Many felt confident in the information they had, crediting doctors, their desire to learn and research, and programs like the annual teen camp where adolescents and doctors gather for a week to educate and offer support to SCD patients.

> "Because over and over again when I come here I am briefed on you know the type of sickle cell and what to do and what the complications are or even the teen camp that was kept, I've been going for five years so I think I have adequate knowledge on sickle cell and what to do and what not do to so would say yes." – *male, 17-19*

> "Well because I will ask my doctors sometimes more about my sickness and stuff and I will like go on internet and research more about the disease so I can get a little bit more knowledge about it so yeah." – *male, 15-16*

3.3.2 Involvement with Disease Treatment and Management

The second round of questions centered on each patient's individual involvement with SCD treatment and management, as well as explaining how confident they felt in managing their health care, and what they would like to do in the future to take care of their health care needs. When asked how important it was to manage their own health, the nature of the response changed as participants got older. Younger participants phrased the level of their importance in a manner that reflected the notion that their parents would not be such an integral part of their lives at some point in the future.

> "It is important to gain as much knowledge as possible because not all the time your parents are going to be around to guide you and to what to eat, what not to eat and how to treat yourself. Your mother or father won't be around to help

> you all the time so you really have to know as much as possible and know how to take care of yourself." – *female, 17-19*

> "It's important because it's not, not, it's not all time your parents will be there for you so you have to know what to do when something happens like if you're just start feeling pain you have to know what to do, you have to make a plan cause not all time your parents will be there. You should have an emergency number, doctor number." – *female, 15-16*

As participants got older, the narrative shifted to personal responsibility in fostering a healthy lifestyle catered to their SCD. One participant, aged 17, understood that there was room for more independence, but indicated that there was still time to take the next steps. However, they were well aware of the responsibilities that awaited.

> "Important, as the saying goes, your health is your responsibility right and nobody can really manage your health for you because only, well only you would know your complications or although a person might have sickle cell disease they might have different complications so […] you'd have to know what works with your body or what eases the pain or what helps in a sense." – *male, 17-19*

Confidence in the ability to manage health care also followed the same pattern. Younger participants, again, framed their responses relative to their parents, saying that they could independently manage themselves, through medication. One participant went on to say that they were able to explain to others what to do in case they fell ill, and their parents were no longer around.

> "I'm confident. […] Because I believe in myself. […] I can fill my own prescriptions, I can take my own medication by myself without mommy. That's how I believe that I'm confident." – *female, 13-14*

Older participants acknowledged changes in their confidence level with time and experience, noting that taking care of oneself requires vigilance and attention.

> "I'm very confident, 100%. […] Because if my mommy should leave today or my daddy should leave today, I know how to take care of myself, I know how to reach the doctor and if somebody don't know how I can explain to them. Even if I'm sick I can explain to them what to do even if I can't help myself I can explain to them what to do and how to help me." – *female, 17-19*

> "Since of lately the confident level has gone up. […] Like you knowing that it's a part of your life know you don't neglect it and just go about your merry way. You have to actually put in the work so that you don't get sick or anything like that, but that's just my opinion." – *male. 17-19*

One participant pointed to limitations in her confidence, indicating the complexity of disease management that also came with independence.

> "I'm okay-ish. To me, knowing that you have a disease and trying to fix it the best that you can is really good so I think that's why I'm good at that." – *female, 17-19*

To gauge reliance on family and strength of networks for support outside the family, the interview included a question on whether the participant had anyone to help with their SCD if their family was not available. Answers for participants were generally friends from school, friends of family, and teachers. One participant, age 18, hinted at a significant other that was not yet serious. However, one participant was clear that there was no one outside his family to whom they could turn, if necessary.

> "No. No one. […] I would take care of myself. I will fight and take it for myself.[…] Cause I just don't keep friend. […] I'm really [cooling] off just here by myself and stuff, so don't really keep friends." – *male, 17-19*

Additional questions to measure independence also asked about booking and making an appointment, as well as their how involved they are during doctor's visits and how much participation they have in any conversation about their health care. Keeping track of and making appointments were usually joint efforts by the participants

and their parents. Older participants, although a few indicated that they had seen the doctor alone, indicated some level of participation by a family member. One participant said they alternated between "public transportation and their aunt." All younger participants visited the clinic with their parents or another adult family member such as an aunt, cousin, or sibling. Many revealed that they assisted their parents in keeping track of appointments.

> "Well when I know when it's time for me to see my, when I have my appointment, I make sure I tell my parents that I have an appointment and they should make sure that I go to the doctor and make sure I get my medication." – *female, 15-16*

> "I know that is like every six months I'm supposed to attend the sickle cell clinic so like I always check it, every six months so my mommy's always there to check it and if she doesn't remember I ask like mommy does the appointment day gone already and she check our chart." – *female, 13-14*

One participant, age 18, did not mention his family in his response.

> "I'm really bad at that. […] By myself. I was supposed to be here from like July the 14th and it's August 14th so I'm really bad at that. It wasn't my fault I just had the fourteen in my head I didn't know which month it was. I guess I'm going to put reminders in my phone now." – *male, 17-19*

A majority of the participants who took part in the interviews could not identify specialized doctors they would see as adults. Only two, a male and a female, both in the 17-19 age group were able to give an answer. The female however, did not relate her response to SCD health care needs, but rather reproductive health care needs.

> "Because I'm a girl and because I'm going to have children in the future I'm supposed to go to the gynecologist and we're supposed to go to normal like checkup doctors, but I'll still come here because I have to." – *female, 17-19*

> "What are the kinds of doctors, well I go to a heart doctor. I don't remember the name. I [also] go to an ophthalmologist, eye doctor right?" – *male, 17-19*

All participants were asked to explain how they would like to manage their health care needs in the future, identifying practices they would like to add or improve. A common response was to become better with adhering to medication and to continue with regular appointments. However, the responses were diverse among everyone in all age groups. Younger participants elaborated on practices for self-care.

> "Checkup often, eat my, keep on eating my vegetables, do my exercise as I get older and older, don't let the bones get week, so. Yeah, that's just, I seem to keep healthy and some." – *male, 15-16*

The most unique answer combined a major step towards curing their SCD, as well as philanthropy.

> "Maybe in the future I would like a bone marrow transplant. […] And actually help who can't afford to do it, try to help them to get one too." – *female, 17-19*

Those ages 17-19 specifically identified the need for insurance and financial security.

> "I mean I going to try my best to remain healthy but in the long run everything takes money so I guess I will have to be working so good money especially if I would have health insurance so getting a good job I suppose." – *male, 17-19*

Finally, participants were asked about how much they took part in the discussion about health care both during doctor's visits, by seeing if they had spent time alone with their providers, and at home. Younger participants generally had not spent time alone. Some showed signs of frustration in the arrangement, saying that the provider would normally speak to her, "but mommy answer[s]." One participant indicated they paid

close attention to the discussions about them, but did not weigh in, while another said providers did not ask as many questions when alone in the room.

> "I sit down, act like I'm not paying attention, but I'm listening." – *female, 13-14*

> Yeah sometimes [I'm alone] but not for long. […] They don't really ask much question as when your mom came in and so on." – *female, 13-14*

When participants were older, they mentioned that providers took privacy into account, and asked parents to leave the room. Time also was a factor, as there was opportunity to foster relationships with doctors.

> "Yeah, when I was going through puberty and she was asking me questions so she asked my parents to step outside." – *female, 17-19*

> "Well, I know my doctors right. I've been here a really long time so I no longer like get nervous for you know appointments as I say before, but the doctors here are really friendly so they would like give off that energy where hey you know don't think of it as a doctor/patient appointment more think of it as like you're speaking to you know just a normal person about your sickness or about what's happening to you today." – *male, 17-19*

3.3.3 Patient Experiences

Part 3 of the in-depth interview focused on personal feelings and experiences with SCD, with their families, and as a patient at the Sickle Cell Unit. Most responses to the questions were positive, with few reservations about doctors and how they felt about their involvement with health care. The section began with the worries they felt about a future with SCD. A majority of participants highlighted their future career options as a source of anxiety, especially those in the 17-19 age group.

> "Well when I was little I wanted to be a pilot but I found out because I have sickle cell and the air pressure up there is not good with us, so I couldn't be a pilot so I changed it to an anesthesiologist and I can be an anesthesiologist

> because I'm not in the airplane. That's the only thing that bothered me because I really wanted to be a pilot. […] But like I'm at my job and I suddenly get sick and then I have to go to the hospital and my boss don't know anything. I'll probably have to tell him and when you have an interview, tell him that I have sickle cell and I might not be at work some days, I might have to go to the clinic." – *female, 17-19*

> "[…] okay I'm not going to say your unemployable but you know that kind of limits what you can do and what if my dream was like to become a professional football player then my dreams would be shattered right there and then." – *male, 17-19*

Another similarity among participants was the fear of debilitating pain or early death due to SCD. One participant, however, would not allow it to hinder quality of life.

> "Oh, what worries me the most is like what I'm going to do if it's got really bad. […] If I get pain. What I'm going to do if it's God will? […] I worry that probably I might die too soon or something like that" – *female, 13-14*

> "I don't know like except that sickle cell children don't have like you know big opportunity of [living] or anything but everybody has to die one day so I don't really mind it you know just live my life to the fullest. […] Yeah I'm not going to really let the sickle cell disease stop me from doing anything I want to because I know I can do all things." – *male, 13-14*

A male in the 17-19 age group stated that relationships and family were the main concern. He pointed out that pursuing any serious relationship meant knowing the genotype of any potential partner, and they had to be HbAA so as to make sure there was no possibility of passing on two copies of the S allele.

> "I mean relationships are a hassle because for me to find a girl with AA is very hard […]. I've never gone to a blood test with a girl just to see if she's capable of me knowing but I guess that would be a factor so that's something to think about." – *male, 17-19*

Some participants however felt confident about what the future head for them.

> "I'm not worried. […] Cause I know that I'm going to be a great, I tell my sister like I wanted to be a doctor but teacher said [..] you're going to be the perfect

> doctor for Jamaica and I was like yes thanks Ms. thank you and she would say , I'm very proud of you […] and your mom will be proud of you as well." – *female, 13-14*

All participants felt they could rely on family as a source of support with the trials and complications that may arise in living with SCD, saying that extended family members volunteered to take them to appointments when their parents could not. One participant claimed that their family was well educated about the illness they had, acting as a "guide" if they "slipped." Family were also foundations for comfort in times of pain and sadness.

> "Yeah, they always like comfort me like when I'm in pain or sometime like when I'm feeling sad or depressed about the sickness and they always like be there for me and stuff."- *male, 15-16*

Willingness and desire to become more involved varied heavily among all participants. Some participants felt there was no need for them to become involved, either because they were already very cognizant of their health care, because they didn't feel it was their place yet at their age, or they felt they were overwhelmed with other aspects of their lives to take on more responsibility. Older participants generally did not feel a need to become more involved.

> "Well I'm already involved because if there's any choice to be made [with] my doctor I'm the one who make[s] it and not my mother making it for me so it's my view first and then if she agrees then we go. Just like the Hydroxyurea. That's a medication? I didn't want to take it because I never want […] medication [that] I [don't] like the side effects at all. I never liked the fact that my bone, what is it again? The marrow? It could destroy my bone marrow and I never liked the idea so whether she agreed that I should go on it, I disagreed so I never go on it, so I'm always involved in every step that takes." – *female, 17-19*

A participant in the 13-14 age group felt they had enough to deal with as a student in school. Another in the 15-16 age group said it was the parents' who should remain involved until he became older.

> "'Cause it's going to be stressful and I'm already on the stress because at school I manage to keep up so I don't want to stress it and I have homework and […] test so I don't really want to put anything else on my head." – *female, 13-14*

> "Not really. I think I should leave that to the parents. I don't really want to go, that's for them. […] When I'm getting much older." – *male, 15-16*

Two questions during the interview attempted to understand patients' relationship with their providers, and how comfortable they felt going to them about whatever problems they may encounter. It also attempted to gauge if participants felt that providers understood their daily struggles with SCD. Relationships were mixed, with some patients treating visits like an means to an end, while others stated that they had personal phone numbers to reach their provider for any questions they may have.

> "Not really. […] When I come to doctor it was [to] get stuff and get your prescription and […] [I] don't really talk to her and stuff. Just come on and get everything […]" – *male, 17-19*

> "I have, [doctor's] number, she has mine say if anything you know I'll call her up and […] say hey what can I do in that instance, you know. So it's not only in this environment in the clinics environment that I can speak to doctors, I can speak to them anytime or anywhere with any concern that I have." – *male, 17-19*

Additionally, personal relationships also varied with participants and their providers. One participant claimed that their provider did not pay attention and was overshadowed by the mother, while another expressed a close, warm connection.

> "Because, my mommy's in control of my sickle cell disease and I feel like they don't care what I have to say." – *female, 17-19*

> "Alright, me and my doctor have a nice one, nice [relationship]. One time she gave me, she um, she see [me] she was run[ning] and come hugs me up, kiss me and so I say, that's my son or that's my boy [name]. I say, and stuff. Every time [I] ask for her, she always find the time to come visit, look for me and stuff. […] I really don't know, but she just love me, she just love me." – *male, 15-16*

Most participants expressed that providers must understand their hardships because they devoted time to the study and treatment of SCD.

> "They have to learn it and know the symptoms and what to give to patients and so on so I think they know how harsh it is." – *female, 13-14*

> "Yeah I mean they like study sickle cell so I guess like they know like what I go through like when I'm in pain like they know like what medicines like what cause it so yeah I think that they have, they're knowledgeable enough to know like what I go through." – *male, 15-16*

One participant felt that doctors had a natural intuition to the experiences they were going through, even before they knew it themselves.

> "Because without even like me stating what's wrong, they can well, when somebody's been through what I have I guess it's obvious that you're going through pain. But otherwise then that they can pinpoint and even tell me how I feel even if I didn't know that I felt it at that time. So like they'll bring up like different choices like they are feeling like this or are you feeling like that and then looking at those choices you're like, oh my God I actually feel like that but I didn't know that I felt like that." – *male, 17-19*

All participants did not feel that the Sickle Cell Unit needed any major improvements. Overall, they felt positively about the manner in which the clinic operated.

> "Well I don't think that, they're just great like overall. I don't think that they need to like improve on anything because you know like, I'm fine with this. They're like just good overall, they don't need like to make any more changes or [anything]." – *male, 15-16*

Some suggestions to implement included making learning about sickle cell more "fun" and that sometimes education about the illness could be "boring". In particular, one participant suggested making the annual teen camp longer, and that it had a positive impact on her life outside the clinic.

> "[…] make the [teen] camp longer. […] my bio class at school now when we're learning genetics, I was the best at it. My teacher was confused, she say you read the whole book already, I said no miss I don't even have the book. I just know this cause, you know I go to the camp and they teach us so I was the only one that answered the questions […]." – *female, 17-19*

One commonality among the positive aspects of the clinic were friendliness, approachability, and openness of the persons who worked there, both doctors and nurses. Moreover, doctors were also respectful of their patients and their choices.

> "Well they're friendly and they're kind and they're always like talk to me, ask me how I'm doing, ask me about my sickness and my experience so yeah I mean they're good. I feel comfortable around them. I can talk to them about anything […] they're just great overall. I don't think that they need to improve on anything." – *male, 15-16*

> "[…] they will put our choice first and they don't tell you or demand you or tell you to do something without you agreeing to it or saying you have to do this. They make you make your own choices so." – *female, 17-19*

The nature of the clinic serving all ages also came up as a major positive aspect about being a patient at the Sickle Cell Unit.

> "Well from I was a baby, well the doctors they love me and know me […] growing up." – *female, 13-14*

The interview concluded with a question that allowed participants to explain anything that made them feel good about themselves. Responses centered mostly around aspects of their personality, their family, their hobbies.

> "Well I guess like [my] overall my personality well I know that like I have like good friends and good family members so like I know like they'll always be there for me and I know I just like me overall like I can't really explain […]." – *male, 15-16*

> "What makes me feel good about myself? Okay I'm like a guy that seeks a lot of attention so when I actually get attention it makes me feel good about myself cause I'm really a dramatic person. […] I do plays and stuff like that […]." – *male, 17-19*

Arguably most importantly, however, participant responses shared similar feelings of their determination to overcome hardship.

> "What makes me feel good about myself is that I'm one of a kind, I go through with things that most people don't ever dream about going through and every time I go through them I come out stronger and better than when I wasn't in them so it gives me a lot of wisdom for my age." – *female, 13-14*

> "[…] What makes me feel good about myself is just that you know the mindset that I have and the warrior spirit, you know to overcome the obstacles and not let anything hinder me because of my sickle cell." – *male, 17-19*

> "Everything, the mind set I have, due to sickle cell and people would say oh you have sickle cell you can't do this, I like to prove people wrong. I'm like a go getter. Yes, so once they say I can't do it, I'll prove you wrong and show you that I can do it." – *female, 17-19*

4. Discussion

4.1 Applicability of Transition Readiness Assessment

Applicability of the Transition Readiness Assessment and Disease Knowledge Questionnaires used in this study are determined by concordance with each other, and with the interviews. Disease knowledge questionnaires for patients in Jamaica, particularly in the 17-19 age group, have an average score of 5.81, about 60% lower than the average score at Duke University. Similarly, the disease knowledge section of the Transition Readiness Assessment also portrays an overall 9% lower self-scoring for the responses for participants in the same age group, 26.58 and 30.27 in Jamaica and Duke, respectively.

In the interviews, there were two questions that received almost unanimous answers. Participants were asked if they knew about the doctors they would see as an adult, and most were not able to articulate a response. A similar question appears on the Transition Readiness Assessment, only asking if they know what is a hematologist. The mean score for this question is 1.72, somewhere between "no, I do not know" and "no, but I'm learning to do this". Second, participants were asked how they attended their doctor's appointments, and if they ever went alone. On the appointment section of the Transition Readiness Assessment, there is also a statement which asks whether the respondent arranges their own transportation. Most participants stated that they had a

family member assist them in making their appointments. The average score for this question is 1.78, also between "no, I do not know" and no, but I'm learning to do this".

Both the Transition Readiness Assessment and disease knowledge questionnaire comparison with Duke, and interviews demonstrate that a standardized assessment is able to be used in Jamaica. Moreover, an assessment similar to those that were used for purposes of this study may able to be used in an international setting when appropriately tailored to the transition needs of an SCD adolescent population.

4.2 The Need for a Transition Program

The Sickle Cell Unit's framework for SCD care from infancy into late adulthood not only provides an opportunity to explore how the absence of a gap in pediatric and adult care affects various aspects of independence in management of health care, but it also can help determine a need for a transition in an area where the concept has not fully developed. Indeed, there are several advantages in remaining with the same doctors, nurses, and staff, as indicated in the interviews. However, when comparing test scores with patients at Duke University, there appears to be serious lapses in disease knowledge and independence that can be gained from a formal transition program.

Jamaican participants ages 17-19 showed improvement in their Transition Readiness Assessment scores when compared to those ages 13-14, particularly in disease knowledge, appointments, and privacy information. The goal of this assessment is to identify gaps in knowledge and readiness in different areas of disease management so

that education, in the form of an area and disease specific transition program, can move all responses into the "yes, I always do this when I need to" category, indicating the patient is ready for transition to adult care. However, all Transition Readiness Assessment scores are consistently lower than their Duke counterparts, and fail to reach maximum potential scores in the appointments section. This may be partially explained by the fact that the measures were meant for use in the United States, but can still indicate an opportunity for intervention in patient transition.

There are efforts to streamline an increased sense of disease knowledge and independence through programs such as the teen camp, a week-long event in the summer that lasts a several hours each day, providing a support system, disease education, and resources for career development. Participants all felt that they had friends to confide in about SCD; however, interviews with participants, even among the 17-19 age group, still indicate that there is hesitation in involvement with health care and potential career paths.

There are certainly strengths to receiving care from the same staff. Participants, if attending doctor's appointments regularly at the clinic for a prolonged period of time, experience a close and intimate and connection with their providers. The level of trust by being able to reach out to doctors at any moment using personal telephones is an asset that, if harnessed correctly, can be a powerful tool in adolescent transition.

4.3 Strengths, Limitations, and Implications for Research

This study is a preliminary investigation on the need for a transition program in Jamaica. There are many strengths and limitations. Strengths include the compilation of current data on disease knowledge, both qualitatively and quantitatively, with interviews providing a narrative to accompany the scores on both assessments. The results show that despite the lack of a gap in pediatric and adult care at this clinic, and even with increasing age, there is considerable variability in all aspects of disease knowledge and independence, and some patients are not well equipped to handle and manage their health care.

A significant weakness is the lack of multiple study sites. The Sickle Cell Unit's comprehensive approach to SCD is unique, and it would be worthwhile to explore if the same patterns exist among patients in Jamaica who frequent public hospitals and clinics. Additionally, similar interviews should be conducted at Duke to understand and discover if their overall increased scores is parallel to the potential responses. Other weaknesses include study sample size and age cutoff. Interviews show evidence that confidence with each topic discussed increases with age, however, there is inconsistency in the levels of their understanding. Expanding this study and recruiting more participants, as well as increasing the age to 21 will not only provide more insight, but will statistically decrease standard error, as well as uncover if a level of stable understanding is reached among all participants in a specified age group.

4.4 Future Implications for Practice

The study aim of assessing transition and readiness for independence was met for the population in Jamaica. It is evident that lifetime care, which is the central aspect of the framework at the Sickle Cell Unit, may not entirely educate and prepare an adolescent patient to become a successfully independent adult with SCD. Further information should be gathered to understand the cultural context and develop tools similar to the Transition Readiness Assessment and Disease Knowledge Questionnaires. However, it may be beneficial to introduce a program that is a consistent part of regular physician visits at an appropriate age where transition can begin to supplement the personal care and attention that is praised as an asset of the clinic. Periodic assessments should be implemented to track results and evaluate the program. In the future, more data should be collected to monitor improvement and incorporate results into a dynamic transition program fit for the needs of adolescents with SCD in Jamaica.

5. Conclusion

Sickle cell disease is the most common Mendelian disorder, and a chronic disease that affects millions globally. While there is definitely a need for research on clinical therapeutic interventions, the consequences of an unsuccessful treatment signifies the transition from pediatric care to adult care is becoming an increasingly important issue. Optimal health care where each person receives developmentally appropriate care results in lifelong functioning and wellbeing for youth.[2] This study provides a preliminary look into the need for a structured transition program in Jamaica, as well as identified patient perspectives and the assets that are unique to the UWI Sickle Cell Unit.

This study takes the first steps in the data collection and furthermore the creation of transition assessment tool developed for patients in a specific country context. These assessments used in this study may serve as a base for similar tools unique to Jamaica. Interviews also provided a unique look into adolescent patient views about independence and their ability to transition, and responses can be used specifically to improve conditions in the clinic. Future studies should expand on the results in several clinic settings, both rural and urban, and public and private. When enough data are

[2] American Academy of Pediatrics. "Supporting the Health Care Transition From Adolescence to Adulthood in the Medical Home." 2011. *Pediatrics* 128 (1): 182–200.

available, a policy can be created for implementation of a culturally appropriate transition program for patients with SCD in Jamaica.

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