

A Whole New World: The experiences of adolescents with beta-thalassemia major as they transition
to adult care

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*It is much more important to know what sort of a patient has a disease than
what sort of a disease a patient has.
~ William Osler*

*The only source of knowledge is experience.
~ Albert Einstein*

*Whenever you find yourself doubting how far you can go; just remember how
far you have come. Remember everything you have faced, all the battles you
have won, and all the fears you have overcome.
~ Unknown*

Abstract

Beta-thalassemia Major (b-TM) is a chronic medical condition. Effective illness management requires adherence to arduous medical treatment to ensure a life free of life-threatening complications. This qualitative study characterizes the experiences of adolescents living with, and managing b-TM on the cusp of transitioning from pediatric to adult orientated institutions of care. Using the methodological approach of interpretive phenomenology and conducting semi-structured interviews with five participants, five themes were identified. These include: knowledge and understanding of b-TM; role of familial support; self-reflection and disclosure; comradery; and readiness to transition, including the impact of incorporating a *Health Transition Specialist* as a member of a comprehensive health care team. The findings identify key factors that impact the daily lives of adolescents with b-TM, as they adapt to required self-management practices, navigate the health care system, and transition to adult care.

Key Words: adolescent; chronic illness; beta-thalassemia major; transition; autonomy

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Chapter One

1.0 Introduction to the Study

The thalasseмии are a group of inherited blood disorders that cause abnormalities in the production of hemoglobin (Cappellini, Cohen, Eleftheriou, Piga, Porter & Taher, 2008). It has been estimated by the World Health Organization (1989, 1994) that globally, over 270 million people are considered “carriers” of the condition, with “... more than 300, 000 children born each year with one of the thalassemia syndromes or one of the structural hemoglobin variants” (Cappellini, Cohen, Porter. et al., 2014, p. 1060). The thalassemia disorders are primarily found in the Mediterranean, Asian, Indian, and Middle Eastern regions (Colah, Gorakshakar & Nadkarni, 2010). The epidemiology of thalassemia, however, is rapidly changing due to migration patterns (*Guidelines for the Clinical Care of Patients with Thalassemia in Canada*, 2009). For instance, Canada has seen an increase in the number of individuals treated for thalassemia due to immigration from countries where thalassemia is prevalent (*Guidelines for the Clinical Care of Patients with Thalassemia in Canada*, 2009).

This thesis explores the experiences of adolescents with beta-thalassemia major (b-TM) as they transition from pediatric to adult institutions of care. An increased life expectancy for those inflicted with the condition is due to the advancements in the treatment and management of thalassemia, which have improved life expectancy well into adulthood (Modell et al, 2008; Cappellini, Cohen, Porter. et al., 2014). As such, it has become increasingly imperative to understand the perspectives of those managing the condition to provide the most optimal health outcomes for this population. As suggested by Cappellini, Cohen, Porter. et al. (2014), individuals with b-TM (and their families and caregivers) are forced “... to deal with its demanding and lifelong management, disability, emotional and other psychological reactions which require adaptation and coping, complex as well as difficult

medication regimens (such as the daily subcutaneous infusions of chelating agents in [beta] thalassemia major), difficult life style and social adjustments and the need for specialized multidisciplinary care.” (p. ii). It is essential that adolescents with b-TM be empowered to gain the self-assurance and tools to manage their condition in order to actively become self-advocates for their needs.

1.1 Evolution of the Study

This chapter provides the researcher’s background on the issue, an overview on how adolescence is defined (within the focus of this study), as well as, the history and disease burden of b-TM. A rationale for the study is provided followed by the study’s main research objectives.

1.2 Researcher’s Background

In interpretive research, the purpose of declaring oneself before any discussion is to provide an overview of what the researcher believes about the particular topic under investigation. This declaration ensures preconceptions are explicit and provides a rationale of how they may enter into the inquiry (Lopez & Willis, 2004).

My passion for research among adolescents with beta-thalassemia major (b-TM) stems from my own experiences living with the condition. B-TM is a life long commitment that impacts each and every aspect of an individual’s life. However, during adolescence, developmental ideals sometimes take precedence over the self-management of b-TM. During my own adolescence, I struggled with the burdensome nature of living with b-TM. Each day brought forth new challenges, pertaining to the health of not only myself but of my fellow peers. Through all of my trials and tribulations, I have come to realize that the thing I struggled most with during my own adolescence was a lack of

understanding from non-patients. This lack was not about the nature of my condition nor of the elements of the management of my care. Rather, it was about the physical, social, and psychological toll living with b-TM takes.

I recognize there are an abundance of factors that have the potential to influence adolescent lives as they emerge into adulthood. However, my experiences as one who has struggled with various complications pertaining to having b-TM throughout adolescence, I can attest to how debilitating the physical and psychosocial effects of living with a condition that few others will ever understand. The complications I faced (and continue to face) have not been trivial. Physically, I overcame several bouts of congestive heart failure, a liver failure, osteoporosis, and continue to manage having hypothyroidism and type-one diabetes. Although significant, these complications do seem minor compared to the sociological and psychological hardships I have struggled with. Through all of these hardships, I never once have felt a true understanding of my perspective as a patient from anyone. Many times I found myself in some of the most renowned institutions within North America and Europe, surrounded by capable medical staff and an exceptional supportive network of family and friends, yet feelings of isolation, frustration, and distrust pursued. After all, *how can any individual or system support someone they do not fully comprehend themselves?*

As a patient I have come to appreciate that living with a complicated chronic disease is difficult and filled with countless challenges, full of a lifetime of worry about our own health and an apprehension of those whom we must put our trust in (healthcare providers). The thalassemia patient population is small, which contributes to the ignorance and lack of appreciation of what we go through. I cannot help but contemplate about what the future will hold. To an outsider, these issues might appear to be peculiar or depressing, but for us, they are consequential.

Reflecting on my past, I can now appreciate that it would be difficult for non-patients to imagine the fears or frustrations of having thalassemia. Furthermore, it may be difficult to empathize with the losses we have experienced, the adversities we continuously push through, or the dreams for a better future we hold on to each day.

I have openly acknowledged these feelings and thoughts to consciously attend to how they may have influenced my interpretation of the experiences of adolescents with b-TM. In doing so, I have attempted to set aside my experiences and beliefs about adolescents living with b-TM, so that I can be open to the meanings that emerged throughout the analytical process. However, one cannot be naïve to appreciate that an individual's knowledge and experiences are never truly absent from the interpretations and analysis of data. I wholeheartedly acknowledge that the findings of this study represent a combination of the meanings articulated by the participants and myself within the focus of the study. And it is my hope that this study contributes to an enhanced understanding of the experiences those inflected with b-TM may face; creating the potential for improvement in the comprehensive care of all individuals living with thalassemia.

1.3 Background

1.3.1 What is thalassemia?

Each of the body's hemoglobin molecules is made up of four proteins (or "globin-chains", typically two alpha and two beta). Each molecule contains a *heme* group containing iron (Weatherall, 2010; Cappellini, Cohen, Eleftheriou, Piga, Porter & Taher, 2008). The various forms of thalassemia have been identified and classified according to the globin-chain they affect (Weatherall, 2010). Classifications of the thalassemias can vary. One manner in which they can be described is by their genetic variations. That is, classification is dependent on which protein globin chain within an

individual's hemoglobin is inadequately produced (Weatherall, 2010). For example, alpha (α) or beta (β) thalassemia or β -chain variants, including Hb E β -thalassemia (Weatherall, 2010). Clinical classifications often focus on the severity of the condition. It is accepted that all clinical forms of thalassemia fall into one of three broad categories: thalassemia minor, thalassemia intermedia, or thalassemia major (Galanello & Origa, 2010).

Individuals with *thalassemia minor* have only one gene mutation and are known as “carriers” of the disease. This type of thalassemia results in no anemia or only slight anemia (Cooley's Anemia Foundation, 2015). Individuals who merely have the mutation do not require blood transfusions. Those whom have inherited one of the more severe thalassemic genes in combination with a milder gene will have *thalassemia intermedia* (Thalassemia Foundation of Canada, 2015). Many of these individuals are able to manage without regular blood transfusions but remain somewhat anemic. *Thalassemia major* (also known as, Beta-thalassemia, Mediterranean anemia or Cooley's anemia) is the most severe form of the thalassemiias. Children are typically diagnosed within the first year of life (Porter, Evangelis, & El-beshlawy, 2011). In these patients, red blood cells are broken down “... almost as soon as they are produced and the bone marrow cannot sustain the demand to produce a sufficient number to replace them...” therefore necessitating the requirement for regular red blood cell transfusions (Thalassemia Foundation of Canada, 2015). Left untreated, the condition will result in severely anemic individuals, whose bodies try to compensate with the expansion of the bone marrow in an effort to produce more red blood cells. This prolonged anemia will cause growth retardation and deformities and eventually lead to death within the first decade of life (Weatherall, 2010b).

Although blood transfusions allow patients to grow normally and to be active, they do not come without costs. One of the consequences of the blood transfusions is the accumulation of excess

iron or *iron overload*, which cannot be excreted naturally by the body (Payne et al, 2008). In order to counteract the potential outcome of organ damage caused by iron overload, patient survival is dependent on adherence to arduous daily chelation therapy. Chelation therapy allows for the removal of excess iron and associated toxicity by achieving and maintaining acceptable iron levels within the body's organs and tissues (Kontoghiorghes, 2009). As you may imagine, the prognosis of these individuals is highly dependent on adherence to the difficult long-term treatment programs, namely the regular blood transfusions, diagnostic testing, and lifelong iron chelation therapy.

1.3.2 Clinical Management of b-TM

With advancements in screening programs, non-invasive iron measurement tools, improved methods of assessing iron burden in the body, safer blood collection and transfusion mechanisms and new drug treatment options, individuals with thalassemia are surviving into their fifth decade and beyond (Modell, Khan, Westwood, Ingram, & Pennell, 2008). However, optimal survival rates and quality of life depend significantly on a comprehensive health care strategy, as well as, adherence to medical review guidelines and medication regimes. This equates to a need for adolescent individuals with b-TM to acquire the skills and understanding to coordinate their medical care, oversee adherence to their treatment, and be able to identify and advocate for any needs that may arise as navigate through their daily lives.

Individuals with b-TM, receive regular red blood cell transfusions every two to five weeks. The amount of blood each person receives is based on an individual's weight and pre-transfusion hemoglobin levels (Cappellini, Cohen, & Eleftheriou, 2008). Regular blood transfusions can prevent the majority of the "serious growth, skeletal, and neurological complications" of thalassemia major (Porter & Viprakasit, 2014). However, transfusion-related complications are considered to be a major source of morbidity among those with b-TM (Porter, Evangelis, & El-Beshlawy, 2011).

As a result of chronic blood transfusions, excess iron stores in the body's organs resulting in *iron overload*. Individuals who are given regular blood transfusions gain a significant increase in unnecessary iron because the human body has no effective mechanism to remove it (Porter & Viprakasit, 2014). The accumulation of excess iron can become lethal, triggering cardiac failure, cirrhosis, liver cancer, growth retardation and various endocrine complications (Porter & Viprakasit, 2014). Therefore, chelation therapy is necessary to remove the potentially harmful excess iron.

Chelation therapy allows accumulated iron from blood transfusions to be removed from the body by increasing iron excretion in urine and or feces (Porter & Viprakasit, 2014). There are currently two forms of chelation therapy available to individuals with b-TM, those administered by intravenous or subcutaneous infusion and/ or those administered by mouth (oral medications). Regardless of its form, every chelating agent has the potential to cause undesirable side effects. These can include, skin reactions, predisposition to infections, severe allergy, divalent ion deficiency (e.g., zinc) and dose-related complications (e.g. toxicity can cause auditory issues, growth retardation, skeletal changes, renal complications, etc.) (Guidelines for the Clinical Care of Patients with Thalassemia in Canada, 2009). Therefore, the daily regimen of chelation therapy requires a significant commitment and individualized coping strategies (Porter, Evangelis, & El-Beshlawy, 2011).

In addition to monthly *Complete Blood Counts (CBC)*, frequent chemistry panels (including *ferritin* assessments), and being seen regularly by a comprehensive hematology healthcare team, individuals with b-TM are often referred to other specialists as a preventive measure or for any issues that might arise due to complications stemming from the condition (e.g. toxicity from chelation medications, blood transfusion reactions, iron overload, etc.). In 2009, inspired by the already established "*Standards for the Clinical Care of Children and Adults with Thalassaemia in the UK*", a dedicated team of healthcare professionals and advocates for compressive care created the

“Guidelines for the Clinical Care of Patients with Thalassemia in Canada” (2009). Below, Figure 1. outlines the recommended monitoring schedule for complications associated with b-TM outlined within the guidebook (*Guidelines for the Clinical Care of Patients with Thalassemia in Canada*, 2009).

Figure 1. Recommended Monitoring for Complications of beta-thalassemia

System	Investigation	Age to start	Frequency of Monitoring
Iron Load	1. Serum ferritin 2. Liver iron content - Liver biopsy - Liver MR 3. Cardiac iron load	<ul style="list-style-type: none"> • Pre-transfusion initiation • After 10 – 20 transfusions • 10 years 	<ul style="list-style-type: none"> • Every 3 months • Yearly • Every 1 – 2 years
Chelation associated	1. Audiometry 2. Ophthalmological 3. Bivalent ion levels (e.g., zinc, copper) 4. Chelator specific laboratory tests	<ul style="list-style-type: none"> • When chelation starts 	<ul style="list-style-type: none"> • Yearly • Yearly • Yearly • Every 1 week to 3 months
Liver Function	1. Liver enzymes - ALT, AST, ALP, Bilirubin	<ul style="list-style-type: none"> • Pre-transfusion initiation 	<ul style="list-style-type: none"> • Every 3 months
Cardiac Function	1. Physical examination 2. Echocardiogram, MUGA scan, or cardiac MRI T2*	<ul style="list-style-type: none"> • At diagnosis • 10 years 	<ul style="list-style-type: none"> • Every 6 months • Yearly
Endocrine Function	1. Short stature - Standing and sitting height 2. Hypogonadism - Puberty staging 3. Hypothyroidism - TSH 4. Hypoparathyroidism - Calcium - Phosphate 5. Diabetes - Random glucose	<ul style="list-style-type: none"> • At diagnosis • 10 years • 12 years • 12 years • Pre-transfusion 	<ul style="list-style-type: none"> • Every 6 months • Yearly until puberty complete • Yearly • Yearly • Every 6 month
Bone Complications	1. Osteopenia/ Osteoporosis - DEXA scan	<ul style="list-style-type: none"> • 10 years 	<ul style="list-style-type: none"> • Every 1 – 2 years
Infections	1. Hepatitis B sAg, sAb 2. Hepatitis C serology 3. HIV serology	<ul style="list-style-type: none"> • Pre-transfusion initiation 	<ul style="list-style-type: none"> • Every 2 – 3 years

Figure 1. Recommended Monitoring for Complications of beta-thalassemia. (2009). Reprinted from *Guidelines for the Clinical Care of Patients with Thalassemia in Canada*, by Anemia Institute for Research & Education and the Thalassemia Foundation of Canada. Retrieved from http://www.thalassemia.ca/wp-content/uploads/Thalassemia-Guidelines_LR.pdf

1.4 Rationale for the Study

B-TM is a demanding monogenic chronic condition, which requires regular blood transfusions to maintain adequate haemoglobin levels. Unfortunately, one consequence of frequent

blood transfusions is the accumulation of iron or *iron overload*, which the body is unable to excrete naturally. For all individuals born before 1970, the diagnosis of b-TM represented an almost-certain death sentence, even in economically prosperous countries such as Canada. However, as a result of innovative technologies, treatments, and management tools, life expectancy of patients has increased over the past three decades, with many individuals living well into their 50's (Brittenham, et al, 1994; Modell et al, 2009; Weatherall, 2010). These favourable survival outcomes for b-TM patients in North America and Europe are observed in parallel with a lack of understanding about the physical, sociological, and psychological toll in young adults dealing with the disease (Trachtenberg et al., 2012). That is, in order to counteract the potential outcome of organ damage caused by the iron overload, patients must adhere to arduous iron chelation therapy. Furthermore, the long-term effects of non-adherence to chelation therapy can be detrimental; these patients will most likely experience organ failure(s), which is a significant cause of death for this population (Porter, Evangelis, & El-Beshlawy, 2011; Payne, 2008).

Gone are the days when b-TM was considered a fatal childhood disease. However, as the life expectancy of those afflicted has increased, the needs of this particular population have also grown drastically. Patients now face new issues, such as gaining independence, obtaining higher education, having families, and securing employment in conjunction with managing the high multidimensional demands of the condition. Additionally, they are also being confronted by new complications, such as bone disease, infertility, and increased occurrences of psychological distress (Mikelli & Tisiantis, 2004; Shafiee, Nazari, Jorjani, Bahraminia, & Sadeghi-Koupaei, 2014; Politis, 1998; Tisiantis, et al, 1996). The adolescent cohort of patients is at risk for poor compliance and treatment management particularly, as they transition through adolescence to adulthood (Porter, Evangelis, & El-Beshlawy, 2011). This danger may be due to the additional burden of dealing with a complex chronic condition

while still facing the customary developmental challenges associated with emerging into adulthood (Eccles et al., 2003). Although health care professionals' intentions aim to achieve positive future health outcomes through implementing current beneficial health behaviours, adolescent conflicting priorities are often influenced by "in-the-moment" decisions. Thus, adolescents are a patient group "most likely to benefit from effective chelation therapy compliance efforts" (Porter, Evangeli, & El-Beshlawy, 2011) and an established continuity of care model in order to successfully facilitate transition (McManus, 2014; Patterson, Budd, Goetz, & Warwick, 1993) from pediatric institutions of care to those trained to manage the care of adults.

Kralik, Visentin, and van Loon (2006) explain that the transition which adolescents burdened with chronic illness face is not as a response to adjustment, but rather it is a part of a process by which humans acclimate and function when they find themselves in a new environment or circumstance. In order to remain consistent with current literature, this review utilizes a well-known definition of transition, which includes, "the purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from child-centered to adult-oriented health care systems" (Blum, Garrell, & Hodgman, 1993, p. 570). Additionally, ideas about the development of an adolescent are based on the premise that adolescence is a critical developmental period marked by significant biological, psychological, and social changes in an individuals' life, typically experienced between the ages of 10 to 18 years (Arnett, 2006; De Michelis & Ferrari, 2016).

This research intends to gain a deeper understanding of how adolescent b-TM patients experience and manage issues surrounding transiting throughout their adolescence to adulthood, in their everyday lives, by utilizing the methodological approach of interpretative phenomenology. Qualitative research that adds to a greater understanding of individual patient experience is important. The approach of interpretative phenomenology as a qualitative health research method, can improve

the ability for health care providers to efficiently respond to the particular needs and concerns of patient populations with compassion and empathy (Munhall, 2007).

1.5 Research Objectives

The four central questions of this study include: (1) what is the meaning of being labelled a paediatric patient/ and or an adult patient, according to adolescents undergoing transition?; (2) what are the experiences of adolescents living with, and managing b-TM?; (3) what challenges or barriers may adolescents with b-TM anticipate for their futures as adults?; and (4) what recommendations may come to light given the experiences adolescents with b-TM?

1.6 Overview of the Study

This chapter provides the researcher's background on the issues under study, an overview of thalassemia and the period defined as adolescence, as well as, the rationale for the study, and its primary research objectives.

Chapter two reviews the published literature and highlights the current body of knowledge on the developmental challenges associated with managing a chronic condition during adolescence, and it acknowledges the gaps within the literature for b-TM patients.

Chapter three describes the study's methodology. The participants, sample selection and recruitment methods, along with the data collection and the data analysis methods utilized for this study. Ethical considerations will be presented.

Chapter four presents the findings of the study. This chapter describes participant's characteristics followed by a detailed description of the results, including emerging themes that were identified.

Chapter five provides a discussion of the findings. It also outlines practical implications and recommendations for improving the self-management and adherence among adolescents with b-TM, ending with concluding thoughts.

Chapter Two

2.0 Review of the Current Literature

This literature review outlines the themes associated with the topic of study, the search strategy used to conduct a review of the current literature pertaining to the focus of study, and current gaps found from the existing literature. During a preliminary literature search on the implications of chronic disorders among adolescents, as well as, my own understanding (and experiences living with several chronic conditions), the following themes were reviewed: adolescence and transition (both physicality and developmentally), the impact of chronic illness on adolescence, transition from pediatric to adult institutions of care, and the experiences of individuals with b-TM.

2.1 Search Strategy

The PubMed, Ovid SP, MEDLINE, and PsycINFO databases were searched using a strategy developed by the *Health Sciences* librarian at the *University of Ontario Institute of Technology* in collaboration with myself, the author. The search combined a wide-ranging list of relevant subject headlines and text-words for chronic illness (both in general and including specifically thalassemia), healthcare transitions, and the adolescent age group. Additionally, a separate literature search was performed to familiarize the author with qualitative methodological approaches to health care research. No date limits were set for literature pertaining to adolescent development and those that focus on transitional programs for adolescents with chronic illnesses. However, a date restriction was aimed at publications from 2000 onward for literature pertaining to the management, treatment, and quality of life of b-TM patients. This restriction was due to the fact that, with the introduction of oral iron chelators and less invasive treatment options for patients, the burden of living with b-TM has changed considerably since this time. Additionally, only results that were published in English were

reviewed. Additional articles were identified from the reference lists of relevant review articles. Search terms were used both singularly and in conjunction with each other. Considerations of alternative spelling and recognized differences in the name of thalassemia were also taken into account.

Relevant research was independently reviewed and those whose abstract described research ultimately unrelated to my research focus were excluded. The abstracts of the remaining articles were reviewed with reference to the following inclusion criteria: (1) the publication was an evaluative study on quality of life of thalassemia patients; (2) the population studied included adolescents and/or young adults with a chronic illness/condition; (3) the publication included outcomes data on transfer from paediatric to adult care; (4) the article presented information on the implications on adolescent development for those with chronic illnesses; (5) qualitative, quantitative, or mixed-method methodologies were utilized; and (6) relevant information pertaining to the disease burden of the thalassemias and, specifically b-TM were discussed. Abstracts that conclusively did not meet at least one of these criteria were excluded. A total of 1867 research documents were found, duplications were removed, records were then screened by abstracts (and excluded if they did not meet the criteria), articles were then assessed for eligibility, and finally 24 articles were critically appraised independently for the purposes of this literature review.

The literature search provided insight into the varied definitions of successful transition. Various research studies focused on attendance at initial appointments (at adult care institutions), others on the time between pediatric and adult visits, and several on both admission and transfer into adult care. Alternatively, some research included in the review focused on the burden of disease for thalassemia patients, which is vital in appreciating that the experiences of adolescents can be impacted by said burden.

2.2 Adolescence and Transition

Adolescence is a period where an individual transitions from childhood into adulthood. It is usually linked with the physical maturation distinguished by puberty, which is the trademark of the interim period (Stalkind, 2002). This period is characterized by various dramatic and rapid changes experienced by the individual biologically, cognitively, behaviourally, socially, and psychologically (Kralik, Visentin, & van Loon, 2006). Everyone, regardless of culture, ethnicity, or health status, faces a developmental period of maturity between childhood and adulthood. During this time and among all of the, sometimes difficult challenges, adolescents begin to develop the required identity that will serve them into adulthood (Salkind, 2002). Additionally, during this period adolescents are attempting to establish their independence, self-esteem, and are typically being asked to consider important educational and vocational choices (Westwood, Langerak & Fieggen, 2014). Below, *Figure 2*. highlights some of the customary developmental challenges faced during adolescence. For those with b-TM, this stage of life becomes even more complicated by the daily management requirements of a chronic disease and the challenge of an anticipated transfer from pediatric to adult care. Adult care institutions are often designed differently lack the concentrated multi-disciplinary resources customarily found in pediatric settings (Busse, et al., 2007; Peters & Laffel, 2011), and differ in how they address issues surrounding development, patient agency, and familial involvement (Westwood, Langerak & Fieggen, 2014). In addition, current specialized biomedical models often dichotomize the treatment of children and adults so that the transition period from adolescence to adulthood falls outside of the primary focus of both pediatric and adult medicine (Anderson & Wolpert, 2004). The experiences adolescents with b-TM have during this pivotal period may shape their ability to self-manage the complexity of their condition in adulthood.

Figure 2. Challenges faced During Adolescent Development

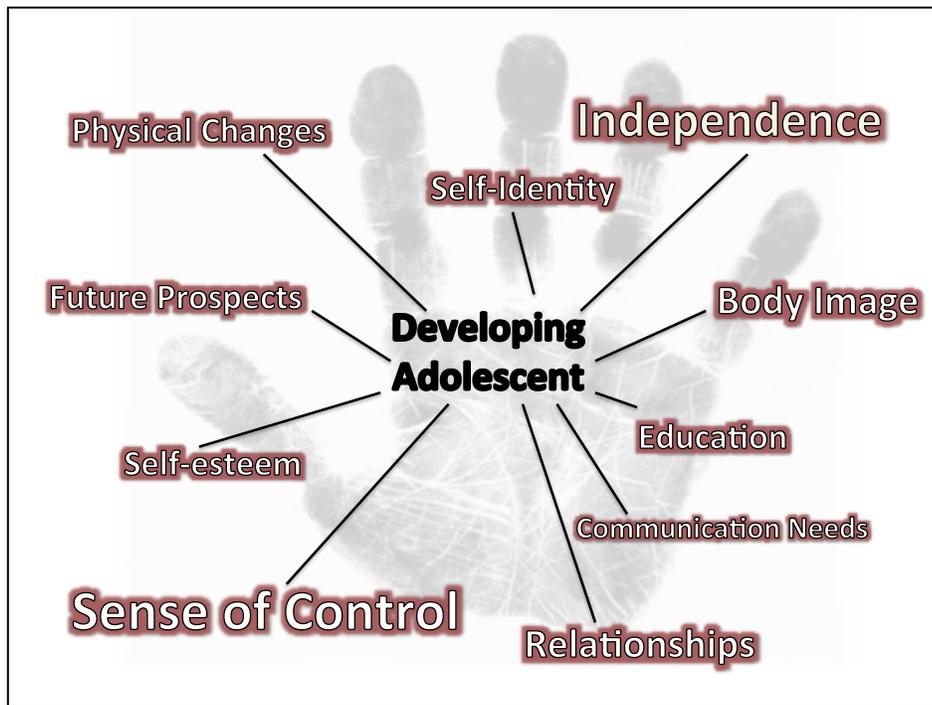


Figure 2. A visual representation of some of the challenges faced by adolescents as they emerge into adulthood. For those with b-TM, this is all taking place in conjunction with managing a complex chronic condition with high stake demands.

For many years, medical transition focused on the physicality of moving a pediatric patient to an adult facility, a relatively simple process. For example, a pediatric individual who requires diabetic care is transferred to an adult endocrinologist’s care, who will continue to monitor the clinical implications of having diabetes. But what happens when a patient’s condition shifts from a fatal childhood diagnosis to a multifaceted chronic condition? Treatment and management for b-TM has never been as promising as it is today, but this has also created new challenges where hematologists face handling the complexities of adolescent development in conjunction with treating b-TM (and any co-morbidities that may arise). Young adults with chronic conditions must develop autonomy in the self-management of disease conditions and learn to communicate effectively with those they depend so much on (their parental guardians and health care team) in order to successfully transition from pediatric to adult oriented health care systems.

2.2.1 The Impact of Chronic Illness on Adolescent Development

A consistent definition of either “chronic illness” or “disability” in pediatrics is difficult to pinpoint, although existing definitions include the concepts of, (a) having particular medical needs; (b) having a clinical diagnosis of a specific mental or physical condition; (c) exhibiting specified functional deficits; and/ or (d) those whom have or are at risk for a chronic physical, developmental, behavioural, or emotional condition which requires related health services beyond the scope of this population generally (Yeo, 2005; Rosen, Blum, Britto, Sawyer, & Siegel, 2003; Steinbeck, Brodie & Towns, 2007). Current literature on the concept of autonomy focuses primarily on older populations, in which the burden and costs associated with chronic disability and illness are tremendously apparent. However the reality is that, chronic conditions, disabilities, developmental progress, and behavioural issues are also prevalent in pediatric populations.

Chronic conditions can impact adolescents in a multitude of ways. In addition to the demands of intricate treatment and management routines, Yeo (2005) suggests that chronic illnesses can affect the rate of young persons’ growth and development, their physical appearance, identity, mental and emotional health, relationships, and enthusiasm concerning education and employment. Furthermore, Lewis and Vitulano (2003) advise that adolescents with chronic conditions can also profoundly impact familial functioning, sibling interactions, and marital relationships. Therefore, it is not hard to appreciate that supporting adolescents with chronic illness can be especially challenging for parents/ guardians and health care providers alike. In comparison to younger pediatric patients, who generally comply with their parental guardians’ requests around health, young adults no longer automatically do what they are instructed. As adolescents’ grow and mature, they learn to become more independent and begin to engage in self-management strategies (Bodenheimer, 2002).

The maturity of self-management practices for adolescents with a chronic condition requires consistent engagement of both parental guardians and adolescents, with attentiveness to the psychosocial realm of the young adult (Bodenheimer, 2002; Watson, 2000). Moreover, Bodenheimer (2002) states that poor family relationships and lower levels of support are associated with decreased adherence rates in young adults, while stronger relationships and connectedness are associated with increased rates of adherence. Therefore, although the facilitation of self-management in adolescents is not a guarantee of adherence, it may be a good start, as lack of understanding and poor commitment to medical treatment regimes are known barriers to adherence in patients.

For adolescents living with chronic illness, additional barriers have been identified in which may increase their risk for a slower rate of achieving developmental milestones. In 2002, Williams, Holmbeck and Greenley conducted a survey of 1434 adolescent patients between the ages of 13-22, throughout six major institutions in the United States. After analyzing the self-reported measures Williams, Holmbeck and Greenley (2002) were able to identify some factors that may impact the development of adolescents with chronic conditions. They included, poor peer relations and/or poor group integration, lack of future planning, difficulty gaining independence, poor or altered body image, dependency on parents and other significant adults, autonomy from parents, absenteeism from school (thus from age-peers), physical restriction, withdrawal, depression, hopelessness, social isolation, delayed social maturation or competence, and affected cognitive skills. Although, Williams, Holmbeck and Greenley (2002) did not distinguish the conditions that their participants were inflicted with, their research is significant because it provides evidence that the development and autonomy needed to effectively manage multifaceted chronic conditions is even more challenged by the complexity of adolescent maturity itself. Therefore, any clinical interventions to aid adolescent

patients must take into account a strong understanding and appreciation of adolescent development, youth participation, and the encouragement of self-management practices.

2.2.2 Transition from Pediatric to Adult Orientated Institutions of Care

In conjunction with the standard biomedical model of care, current literature on the transitioning of adolescents to adult orientated institutions of care focuses on the individual, rather than on the implementation of an innovative health care framework that would be necessary to care for this cohort of individuals. In 2011, Andiman discussed the recommendation of multidisciplinary health care teams in order to facilitate successful transitions. His suggestions also included the development of an appropriate care model, a pre-planned and methodical transition, and the consideration of the psychosocial needs of adolescent populations (Andiman, 2011). This corresponds to the work of earlier researchers, Delengowski and Dugan-Jordan (1986), who shared their personal experiences working with adolescents in an oncology department and detail how specialized education impacted their experiences. That is, the authors stated that a health care team requires the awareness of adolescent development and growth in order to care for these individuals successfully. Schidlow and Fiel (2000) also affirm that the majority of adult orientated health care teams have not been adequately educated to work with adolescent individuals. In addition, Bowen, Henske, and Potter (2010) recognize that the transition of adolescent patients may require additional training for health care professionals and that this training should include “aspects of the developmental, emotional, and social needs of adolescents” (p. 101). Therefore, it is essential that educational and professional development tools be available for all practitioners and staff who will be responsible for the chronic health care needs of adolescent patients as they transition from pediatric to adult centers of care.

In 2011, the *American Academy of Pediatrics* released a publication, which acknowledges that although adult health care professionals are required to assume the care of adolescents, they cannot be expected to be knowledgeable in how to do this without supplementary resources, such as education. The report also states, “Further work is needed to characterize, demonstrate, and teach an adult model of care that is responsive to the particular needs of all young adults and sensitive to the specific challenges associated with providing high quality care to young adults with specific chronic conditions” (Academy of Pediatrics, 2011, p. 197). As the global burden of disease continues to shift from fatal infectious disease to manageable chronic conditions, the transitioning of adolescent patients to adult health care institutions should be of special interest to practitioners and health care staff alike. That is, as conditions, just like b-TM, move from fatal childhood disease status to those where effective management and treatment have the ability to prolong life expectancy, individuals will be dependent of a knowledgeable staff who understands their needs and acknowledges their concerns.

2.3 Experiences of Individuals with Beta-thalassemia Major (b-TM)

There are too many confounding variables to estimate the generalizability of the experience of living with b-TM. This is very evident in the clinical depiction of thalassemia patients’ care. That is, even within the most severe form of the condition, *beta-thalassemia major*, symptoms vary from extremely debilitating to mild in those who are closely monitored, receiving regular red blood cell transfusions, and meticulously observed chelation therapy. In addition to this concern, individuals with b-TM have not been consistently evaluated on how the complexity of living with the condition impacts their personal and professional development, psychosocial wellbeing, and quality of life.

Koutelekos and Haliasos (2013) investigated the relationship between depression and thalassemia in children, adolescents, and adults. By conducting a systematic literature review Koutelekos and Haliasos (2013) acknowledged that the psychological wellbeing of paediatric and adolescent individuals living with b-TM is not fully understood. Furthermore, Koutelekos and Haliasos (2013) were able to conclude that depression among adolescents should be of particular concern to care givers. The researchers go on to state, that as adolescent individuals realize the impact of what their disease burden will entail (throughout their entire lives) on top of the customary challenges associated with this period, the onset of depression is more likely (Koutelekos & Haliasos, 2013). Koutelekos and Haliasos (2013) identified three main areas that may predominantly affect adolescents, these include; (1) family (overprotective, negligent, or hostile parents); (2) social (uncompassionate peers, lack of understanding from non-patients); and (3) burden of disease (blood transfusions, chelation therapy, complications, accumulated or allocated time). In addition to this, the researchers conclude that feeling supported by family, friends, and their health care team all play a critical role in building confidence and independence necessary for managing thalassemia.

Survival for b-TM patients is knowingly dependent on adherence to chelation therapy. Although advancements throughout the past three decades has made this process easier for many patients with the invention of oral chelating medications, not all patients are eligible or are able to tolerate them. For these patients, subcutaneous nightly infusions are the only method to remove excess iron caused by blood transfusions. Regardless of method, iron chelation medications come with a slew of side effects, such as gastrointestinal issues, pain (from needle injections and injection “sites”), rashes, itchiness, as well as the arduous time consumption. Many studies have demonstrated that age is significantly correlated with adherence; where adolescents were shown to have a lower adherence than younger children (Kipps et al., 2002; Williams, Holmbeck, & Greenly, 2002;

Annunziato et al., 2007). According to Porter, Evangelini and El-Beshlawy (2011), lower levels of adherence may be due to several factors, chelation type (side effects dependent on chelator type), psychosocial issues, as a means to affirm independence; and simply, forgetting to take medication. Therefore, being able to identify the factors that influence adherence in adolescents remains an essential first step to facilitating successful health outcomes.

The effect that an increased knowledge of one's condition is associated with an increase in adherence to chelation therapy was investigated in a group of 32 individuals with thalassemia with a mean age 17 years (Lee, Lin, & Tasi, 2009). The results from the cross-sectional correlational survey concluded that there was a positive association between adherence and knowledge. In addition, Lee, Lin, and Tasi (2009) acknowledge the very apparent need for systematic education for individuals with thalassemia, their caregivers, and health care teams to improve adherence to their treatment.

Advancements in the treatment and management of b-TM continue to improve, however individuals with b-TM now face new challenges, some that have not been investigated and others, of which little is known. For example, what the possible impact that living with thalassemia has on educational and employment goals and feasible achievability. Pakbaz et al. (2010) used data obtained in the first longitudinal *Thalassemia Longitudinal Cohort Registry* conducted in North America in 2010 when he investigated the self-reported measures of education and employment in 633 thalassemia patients. Although the registry included all variations of the thalassemia population and a broad age range (5-75 years) it still provided valuable insight on the experiences of thalassemia patients in North America as a whole. Pakbaz et al. (2010) concluded that 47% of those over the age of eighteen (349 patients) were employed full-time, 23% part-time, and 30% were not employed at all. Further, they found that individuals who were transfused and receiving regular chelation were 6.7 times more likely to be employed than those who admitted to being non-adherent to their chelation

therapy. Regular chelation also was found to be a significant predictor on higher education among those with thalassemia, with regular chelation accounting for patients being 3.7 times more likely to have achieved a higher level of education (Pakbaz et al., 2010). The researchers provided a possible hypothesis for the positive relationship between regular chelation and employment/ education rates in individuals who were transfused; suggesting that this may be due to improved health and a reduction in complications compared to those who were not consistently chelated. Furthermore, Pakbaz et al. (2010) inferred that the participants in their investigation might be generally more motivated, as is evident by their efforts to adhere to their chelation therapy.

Also utilizing the *Thalassemia Longitudinal Cohort Registry*, Sobota et al. (2011) investigated the prevalence and incidence of complications in b-TM patients. Using data obtained from the self-reported questionnaires and patient medical records Sobota et al. (2011) were able to provide evidence that females, older patients, and those with more disease specific complications (including side effects from chelation therapy) reported significantly lower quality of life scores. Furthermore, Sobota et al. (2011) suggested that adolescents and adults with thalassemia reported lower quality of life measures than the United States population overall, despite the available advancement in treatment options available to individuals with the condition today.

Similarly, Porter, Evangeli, and El-Beshlawy (2011) note that iron chelation therapy may be responsible for a major decline in the quality of life among individuals with b-TM. Because of this, Porter, Evangeli, and El-Beshlawy (2011) advocate for a multidisciplinary team approach (which should include a clinical psychologist) to optimize adherence results among those with b-TM. Adolescent individuals with b-TM may become more of a challenge because of their desire to express individuality and independence. That is, for some adolescent individuals a “doctor’s orders” to comply with an arduous treatment plan is an opportunity to assert themselves as autonomous (Porter,

Evangelini, & El-Beshlawy, 2011). It then comes as no surprise, that the adolescent aged cohort is the patient group most likely to benefit from effective chelation therapy and transitional intervention (Trachtenberg, Vichinsky, Haines, et al., 2011).

The need for psychological support for those adolescents with chronic conditions is widely accepted (Falvo, 2014; Lubkin 2014), as is the negative effect of psychological issues during this developmental period on chelation adherence in b-TM (Porter, Evangelini, El-Beshlawy, 2011; Evangelini, Mughal, Porter, 2010; Porter & Davis, 2002; Panitz & Sugar, 1999). Unfortunately, after conducting an extensive *Cochrane Review*, Yamashita, Mednick, Haines, and Porter (2014), reported that "... that no randomized controlled trials employing psychological therapies ... were identified" and "no trials, where quasi-randomization methods such as alteration", were found. This is disheartening, given that adolescents' adherence to vital chelation therapy may negatively impact long-term survival rates (Efthimiadis, Hassapopoulou, & Tsikaderis, 2006; Modell, Khan, Darlison, 2000).

One study that was conducted by Roy, Chatterjee, and Chopra (2007) investigates the psychological burdens in an adolescent population living in West Bengal, India. Roy, Chatterjee, and Chopra (2007) suggest that culture and education play a major role in illness experiences. Furthermore, they suggest that the consequence of living with thalassemia is tremendously stressful, and those with the condition face a multitude of physical, psychological, and social problems. However, given the disadvantaged setting (e.g. lack of access to regular blood transfusions and adequate chelation medications), it should be considered that this population might face challenges that vary from those in more developed countries.

Generally, despite a shortage of literature on the psychological impact living with b-TM has on those inflicted, there appears to be some global evidence that psychological well-being impacts adherence to chelation therapy and thus, quality of life.

2.4 Gaps in Current Literature Pertaining to the Study

The body is the essence of body image and self-image, as it provides a perspective which a person identifies and interprets experience. The notion that a sick body is a facilitator of a valuable social existence is significant in the case of adolescents with a chronic illness or condition. Their circumstances not necessarily exclude them from normalcy but rather it is the focus of one's social interpretation. This is why one adapts, to try to reduce the implications of said condition as much as possible. But what happens when that body does not functionally meet the expectations of an individual? Or society? For individuals with thalassemia, attempting to normalize their bodies and their lives is a way to minimize the disruption that managing a multifaceted illness brings. Thus, non-adherence to chelation therapy or the management of their condition may appear as an attitude of defiance or ignorance, but in reality it may be a result of expressing the need to normalize one's body and life, even when the consequences of doing so are detrimental.

As evident from the literature, self-management of chronic illness presumes that individuals with chronic conditions are autonomous adults. However, adolescents do not miraculously develop the ability to be independent on their eighteenth or nineteenth birthday (age of majority across Canada). Rather, maturity comes from the facilitation of patient empowerment –which encompasses the understanding, knowledge, acceptance, and shared-decision making skills that reinforce self-management practices.

Adolescent individuals with b-TM run a particularly high risk of poor adherence and

treatment management as they transition from adolescence to adulthood. This may be due to the additional burden of dealing with a complex chronic condition while still facing the customary developmental challenges associated with emerging into adulthood. The transitional protocol of all health care institutions may fail to ensure that developmentally appropriate services are available and accessible in a continuous and effective manner, as individuals with b-TM move through adolescence to adulthood. The challenge to improve patient outcomes as a result of successful transition should be shared by both pediatric and adult practitioners alike. It is apparent that more research should be explored in order to facilitate the most effective manner to transition adolescent b-TM patients into adulthood by fully comprehending the needs of this particular population.

Chapter Three

3.0 Methods, Data Collection and Analysis

The purpose of this study is to explore the experiences of adolescents' individuals living with b-TM as they transition to adult care. A purposive sampling (Patton, 2001) of adolescents with b-TM, aged 16 to 18 years of age recruited from, the *Hospital for Sick Children* (HSC/ Sick Kids) in Toronto, which is considered a “gold-star” center of care for pediatric hemoglobinopathy patients in Canada.

In this chapter, I describe the study's methodology, design, sample, and sampling procedures. In addition, details are provided on data collection and analysis protocols, as well as, ethical considerations. A phenomenological research design was employed. Furthermore, utilizing a qualitative research allows for the study of a phenomenon in its natural setting. Using a qualitative paradigm allow researchers the opportunity to acquire an idiographic understanding of their participants, and what their experiences mean to them (within their social reality) to live with a particular condition or be in a particular situation (Bryman, 1988; Patton, 2001).

3.1 Ethical Considerations

3.1.1 Research Ethics Board Approval

An ethics board review of this study was approved by both the *Hospital for Sick Children* (aka SickKids - REB file No. 1000056051) and the *University of Ontario Institute of Technology* (REB file No. 14289). A copy of both approval letters can be found in *Appendix A* and *Appendix B* respectively.

3.1.2 Obtaining Consent

All participants were fully informed about the study's objectives and procedures, written consent to participate. In accordance with the *Hospital for Sick Children's* policy, all consent forms remained onsite, located in a secure double-locked location within a member of the research teams office.

3.1.3 Confidentiality and Well-Being

Participants were given the opportunity to stop for a break or to completely withdraw from the study at any time, with no consequences before, during, or after data collection. Participants were also reminded that if at any point they were not comfortable answering a particular question they could decline to answer without question. Because this study involved consenting adolescents (Canadian Institutes of Health Research, Natural Sciences and Engineering Research Council of Canada, and Social Sciences and Humanities Research Council of Canada, 2014) who spoke about their past experiences, two foreseeable risks were considered. Firstly, that participating in the study could lead to the chance of discomfort of recollecting particular experiences of learning to live with, and manage b-TM as participants were asked to provide honest and candid information throughout the interviews. Additionally, I also encouraged participants to elaborate on responses which I felt warranted additional explanation. To minimize any risk of discomfort, participants were reminded that they could choose to leave the study at any time with no explanation and would suffer no negative consequences, including any changes in the care they received at the *Hospital for Sick Children*.

Furthermore, as part of the information letter (participants were given a copy included with their signed consent form), participants also received the contact information for all the members of the research team and their current hemoglobinopathy *Health Transition Specialist* should they have any questions, comments, or concerns about the research study.

Secondly, there was risk that participants' interview data could be identified by members outside their immediate circle of care and/ or their peers. As such, participant data were de-identified and transcribed to maintain participant confidentiality. Raw data were only available to the members of the study's research team. All data were kept on an encrypted external hard drive in a locked filing cabinet when not in use.

Following transcription, participants were asked to review their experiences through the process of member checking. If a participant determined that any of the findings were unrepresentative of their experiences or provided responses, they could amend incorrect interpretations through email correspondence. Any email correspondence was securely deleted from the UOITnet server once it was confirmed that the authenticity of participants' experiences was fully understood. These actions significantly lowered the risk of harm to participants, as they had the opportunity to reach out and seek guidance from members of their immediate circle of care, as well as, confidentiality of their responses and experiences was maintained.

3.2 Research Design

3.2.1 Theoretical Framework

This research is guided by the interpretivist paradigm. Within qualitative inquiry, a paradigm identifies beliefs and principles that guide researchers when considering a study protocol, as well as, aids in establishing a set of practices (Guba & Lincoln, 1994). These practices may include seeking to explore a specific phenomenon of interest -- in this case the experience of being an adolescent living with, and managing, b-TM. This is done through understanding the meanings and intentions people give to their own actions, as well as, their interactions with others (Given, 2008). Daily occurrences

or *lived experiences* that are taken for granted can be examined to better understand meaning and aid in bridging gaps of the understanding of specific phenomena.

In following an interpretivist paradigm throughout my study, I acknowledged that the realities of my participants are multiple, fluid, and collectively co-constructed (Cohen & Crabtree, 2008). As such, reflexivity (on personal, professional, methodological, and contextual level) played an essential role in shaping my interpretations and I made sure to take field notes and keep a journal of my thoughts and perceptions throughout the entire research process. Additionally, in keeping with the perspectives of qualitative inquiry, my approach attempted to understand, as well as, provide a meaningful account of the realities of my participants (Cohen & Crabtree, 2008). The strength and credibility of an interpretivist paradigm lies in the ability to interpret, address the complexity, and find the meaning of everyday occurrences. It is here where my own past experiences as an adolescent learning to manage b-TM are an essential aspect to understanding meaning that may lay under-the-surface of my participant's narratives.

3.2.2 Phenomenology

According to its founder, Edmund Husserl (1970), *phenomenology* is defined as “the science of essence of consciousness” and his focus surrounded the idea of intentionality and the meaning of lived experience. As such, Husserl's phenomenology focused on the question, “*What do we know as persons?*,” where everyday occurrences or experiences are described in rich detail while any preconceived ideas or biases are “bracketed” by the researcher (Reiners, 2012). A fundamental ideology of Husserl's approach to scientific inquiry was the belief that the meaning of “lived experiences” may be understood solely based on one-to-one interactions between a researcher and the entity of research. The researchers interactions must involve attentive listening and observation to construct a depiction of reality more refined than preceding understandings (Husserl, 1970).

To answer the research inquiry of this study, interpretive or hermeneutic phenomenology was utilized as a methodological approach. Martin Heidegger developed hermeneutic phenomenology (the philosophy of interpretation) in opposition to the transcendental phenomenology of Husserl (Reiners, 2012). Heidegger's views called into question the assumptions and accepted traditional thinking of philosophers from Plato to Descartes to Kant (Morton, 2000). Hermeneutics focuses on human experience to uncover what is normally hidden in life experiences. Hermeneutics goes beyond description and seeks meaning embedded in *lived experience* (Lopez & Willis, 2004), enabling interpretation of the world of everyday life through these experiences (Lopez & Willis, 2004; Lynch-Sauer, 1985). Unlike Husserl, Heidegger believed that meaning could be found embedded in everyday occurrences. Thus, the critical question asked by Heidegger is, "*What is being?*" (Reiners, 2012). According to the Heideggerian (1962) traditions of phenomenology, "humans are embedded in their world to such an extent that subjective experiences are linked with social, cultural, and even political contexts." Heidegger focus was on ontological questions of what it means to be a person, of what sort of beings we are, and how this relates to intelligibility (Morton, 2000). This shift was a radical break with traditional philosophical questions concerned with epistemology. Heidegger emphasized social context, believing that most philosophical questions were formed by everyday social practices (Dreyfus, 1991).

From Heidegger's perspective of phenomenology, phenomenology "... provides a guide for the exploration of a phenomenon of interest through the in-depth understanding of a participant's experiences" (Benner, 1994). The meaning constructed from participants' experiences relies on the interpretation or analysis of a researcher (Heidegger, 1962). It is here where I believe my own past, as an adolescent individual living with b-TM, has been essential in interpreting the experiences of my participants.

Interpretive phenomenology hopes to develop understanding and provide “animated and evocative explanations and descriptions based on lived experiences that demonstrate language, traditions, meanings, concerns and practices” (Van Manen, p. 19). For those with a chronic condition as complex as b-TM, this understanding is a part of being able to manage the demands of their condition, and the question is, "what are the experiences of those who face these challenges from their perspective?" This implies that, the experiences of medically vulnerable adolescents who require complex specialized medical care (like those with b-TM) are considered as taken for granted occurrences. The most suited method to reveal the “taken for granted” meaning in everyday experiences among adolescent with b-TM is interpretive phenomenology. Understanding these experiences and meanings will help healthcare practitioners to provide more comprehensive care for adolescents who have b-TM.

3.2.3 Application of Interpretative Phenomenology to Research Question

This study proposes that using a phenomenological approach provides a deeper understanding of the experiences of adolescents, living with, and managing b-TM, on the cusp of transitioning. Utilizing this qualitative approach and semi-structured interview technique will yield data that illustrates the real-life experiences that reflect the developmental dynamic that surrounds the readiness of adolescents' transition to self-management in adulthood.

According to Jopling (1996), phenomenology “... is a critical reflection on conscious experience, rather than subconscious motivation, and is designed to uncover the essential invariant features of that experience ...” (p. 304). Therefore, a phenomenological approach in qualitative research begins with recognizing a phenomenon of interest and realizing the significance of an individual’s subjective experience (Goulding, 2003, p. 304). Using the semi-structured interview method of gathering data, adolescents’ experiences and perceptions of the transition process were

explored. The constant comparative method of data analysis was used to identify and compare viewpoints across individual and group adolescent experiences. Through this process, recurrent themes were identified and connected. From it emerged a deeper understanding of how adolescents are impacted by the process of transitioning (both developmentally and physically).

3.2.4 Criteria for Evaluating Interpretative Phenomenological Research

According to Creswell (2007), qualitative research demands rigorous methods of data collection, reflection, analysis, and writing. Correspondingly, Denzin and Lincoln (2005) suggest that common terminology used in quantitative research such as *internal and external validity, reliability, and objectivity*, are exchanged in qualitative research with the terms *credibility, transferability, dependability, and confirmability*. Credibility in qualitative research aims for findings to offer an accurate and valid portrayal of an individual's lived experience (Lincoln & Guba, 1985). In this study various methods were used to improve the credibility of the study, including member checking. This process involves a review of participant transcripts, as well as, summarizing resultant themes for consenting participants and requesting that they confirm that the themes accurately represented their experience. Additionally, post-interview conversations engaged all participants in collaborative reflection and validation of emerging themes. Participants who indicated that their experiences were misrepresented were asked to provide additional details to guide in reanalyzing any identified inconsistencies (via email). This step was an effective way to ensure that findings were accurate and valid (Rolfe, 2004).

Transferability lies within the researcher's interpretations as to whether or not the findings apply to other settings (Lincoln & Guba, 1985). To allow myself ample information from which to infer their findings, the overall study's objectives were disclosed to participants in detail prior to the initial interview. Additionally, discussed in *Chapter Five* (section 5.5) the transferability of how this

study's inquiry may be transferable to other contexts, including suggestions of how they might or might not apply to other settings or populations.

Dependability in qualitative research refers to the reliability, or repeatability, of the findings within a study (Lincoln & Guba, 1985). In this particular research initiative, dependability was safeguarded using an inquiry audit. Utilizing my understanding of participant experiences, as well as reviewing interpretations with professional experts facilitated logical explanations throughout the thematic analysis.

Confirmability is the attempt to discount researcher subjectivity based on biases (Lincoln & Guba, 1985). Throughout the data collection process and analysis, I maintained an audit trail to provide a detailed outline of the conscious and physical steps taken. An audit trail establishes how theme identification arises from an individual transcript and questionnaire, to demonstrate how themes are derived from the findings (Reid & Gough, 2000).

3.3 Setting and Participants

3.3.1 The Hospital for Sick Children (Sick Kids)

Approximately 180 individuals are treated for thalassemia at the *Hospital for Sick Children* in Toronto, these young and adolescent individuals make routine visits to receive treatment and be seen by a comprehensive hemoglobinopathy health care team as needed.

According to their website, *The Hospital for Sick Children* “is Canada's most research-intensive hospital” and delivers the most innovative and comprehensive care in the “... largest centre dedicated to improving children's health in the country” (“About SickKids”, 2018). Before any ethics application was considered, the hospital required that I join their organization as a student researcher (which required attending an orientation and training), as well as, sign a confidentiality agreement

and adhere to their code of conduct while conducting my research on their premises. I had an assigned to a research supervisor in the hematology department who supervised my research and reviewed all documentation required by the *Research Ethics Board*. Additionally, I worked closely with the current *Health Transition Specialist* to enroll participants, as well as, design and implement the study.

3.3.2 Health Transition Specialist

Although treatment for those living with b-TM remains generally consistent, as adolescents gain independence they must confront a variety of new challenges as they emerge into adulthood, such as transferring out of pediatric care, moving away from home, higher education and/ or employment, and family planning. Innovatively, in 2014 the *Hospital for Sick Children* created a specialized role for a *Health Transition Specialist*, whose role includes supporting adolescents and young adults with red blood cell disorders by aiding in the preparation for and throughout the transition from pediatric to adult health care.

Interestingly, after a systematic literature review on “transitional navigators” very little consensus appears to be reached on the role and definition of a transitional coordinator. The review revealed, of ten studies that evaluated health outcomes following transition programs – only three had included or referred to a “transition coordinator, transitional navigator, or case manager” (only two of which were considered successful programs) – the term *case manager* was also used analogously (Crowley et al, 2011). Furthermore, the role of *transitional coordinators/ transitional navigators/ case managers* was ambiguous (Van Wallegghem, MacDonald, & Dean, 2008; Grant, & Pan, 2011; Kingsnorth, Gall, Beayni, & Rigby, 2011).

According to the hemoglobinopathy team I worked closely with at the *Hospital for Sick Children*, this role was implemented within the clinic mainly to assess adolescents’ and young adults’

readiness for transfer and knowledge of health condition at multiple time points; to provide tailored education, guidance and support to adolescents' in developing self-management skills; and to facilitate smooth transition between pediatric and the adult institution of care.

3.3.3 Inclusion/ Exclusion Criteria

The inclusion criteria of my proposed participants are as follows: (1) a clinical diagnosis of b-TM, (2) participants should be receiving the majority (at least 75%) acute care at the HSC and (3) participants should be between the ages of sixteen and eighteen. The exclusion criteria for research participants are patients: (1) who receive more than 75% of their acute care at other hospitals or “satellite clinics” (community-based primary care clinics), (2) who do not speak English, and (3) who are over the age of eighteen (as these participants would have had a high probability of already utilizing adult institutions for any “emergency” visits and thus have been exposed to adult disease management models).

3.3.4 Recruitment

All eligible patients were identified and first contacted by the hemoglobinopathy *Health Transition Specialist*, and who expressed their willingness to participate in the research study. Those who expressed interest were then formally introduced to myself on an already scheduled clinic appointment date, official consent was obtained (through the signature of a consent form), and basic demographic and interview data were collected. Adolescent participants self-identified as having first-hand knowledge of the experience of living with b-TM, and had the ability to reflect on and articulate their experiences.

This recruitment strategy was recommended by the *Ethics Review Board* at the hospital in order to adhere to the policy of having a member of individuals immediate “centre of care” introduce any study that they may be eligible for when possible. When a possible participant expressed interest

in the study and the *Health Transition Specialist* confirmed their eligibility, I was sent an email that notified me of their name and next clinic appointment date. As per hospital protocol, a private meeting room was arranged through a nurse coordinator of this clinic.

3.4 Data Collection

3.4.1 Instruments and Interviews

A *Thalassemia Readiness Survey* (see *Appendix C*) was administered to each participant, merely to collect descriptive data only. This survey was adapted by the current *Health Transition Specialist*, from the *Hospital for Sick Children's* "Good to Go" general adolescent transitioning program (this questionnaire has typically been administered to those patients who will begin receiving their treatments at an adult institution within approximately three years). Additionally, socio-demographic and clinical information was collected at the time of an initial interview through the self-reporting questionnaire. For each patient, the following data was collected: age, gender, iron chelation therapy type, self-reported measure of iron burden, and average schedule of red blood cell transfusion treatments at the HSC. After participants completed the questionnaire (which took an average of 10 minutes), they were interviewed regarding their experiences as an adolescent patient, their readiness to transition to an adult care facility, any concerns or anxieties they may have about transitioning, and recommendations they felt may help them make their journeys' more successful (see *Appendix D* for proposed interview questions). These semi-structured interviews lasted an average of seventy-five minutes. Interview sessions were recorded on and field notes were documented for each individual participant, as well as, my own reflections. After each interview was transcribed, the audio recordings on an encrypted USB drive were permanently deleted. After meeting to complete the initial questionnaire and interview, participants were asked to provide an

email address that they could be contacted at should I require further clarification of any part of their interview and as a means to review a transcript of their experiences in order to ascertain the accuracy of my interpretations.

3.5 Data Analysis

Each of the participants were given a pseudonym and all other names or identifying pieces of information were altered or eliminated to maintain the privacy and confidentiality of all individuals. With the exception of identifiers, all interviews were transcribed verbatim. After verifying the meaning, context, and understanding of my participants' initial interviews with them, transcripts were uploaded to NVivo 11 – Pro Edition (version 11.4.1.1064 (64 bit)) to aid in the interpretive and thematic analysis. This process also enabled overall data collection and analysis to occur at the same time (Idczak, 2007).

Constantly re-reading through each participant interview while simultaneously returning to the literature (pertaining both to adolescent development, as well as, transitioning while living with a chronic illness) established an overview (Halldorsdottir, 2000) of the lived experience of adolescents living with, and managing b-TM. Additionally, key experiences, explanations, or interpretations were recognized and highlighted by myself. As similar emerging themes were recognized between participant interviews they were grouped and categorized within NVivo for easier comparison and interpretation. In order to help facilitate this process, and prior to data analysis I enrolled in and successfully completed an online course (*Fundamentals of NVivo 11*, February 20 – March 20, 2017), which aided the evaluation of my participant data.

After transcripts were uploaded into NVivo, I coded the data. Assigning themes to data is considered “coding” (Yin, 2016). I began coding with themes that appeared most explicit (assigning

each “theme” a specific highlighted colour) and evolved into creating themes that were less apparent on the surface. Excerpts encompassing themes were compiled into a table, which exhibited how ideas and experiences were linked. Ultimately, the analysis involves synthesizing identified themes with current literature. It is significant to note that although the steps outlined above appear to be singular steps within the analytical process, this is not the case. Rather, the process is a continuous cycle of repetitive phases.

Also, confirming my understanding of participant experiences, as well as, reviewing interpretations with my supervisor, committee members, and research team members from the *Hospital for Sick Children*, facilitated logical explanations during thematic analysis (Houghton, Casey, Shaw, & Murphy, 2013). This review was an additional safeguard in allowing optimal interpretation of my participants’ experiences and minimized the risk of misinterpretation.

3.5.2 Establishing Trustworthiness (Reflexivity and Validity)

My own intimate experience with and knowledge of living with and managing b-TM as an adolescent permitted me the benefit of creating a foundation for establishing trustworthiness. As a part of the relationship building process encouraged when initiating semi-structured interviewing and after suggestions from my hospital supervisor, I disclosed my own status as a “fellow patient” during my participant interviews. Although, I imagined that my mention of a health-related connection was expected to initiate participants' appreciation that my interest goes beyond a scientific agenda and is informed by a personal-experienced based appreciation, I was significantly astonished by the richness of data I collected after my disclosure. Participants recognized that my sharing with them that I have b-TM, as it was intended to, fostered a sense of assurance that I intended to treat them and the information they shared in a respectful and trustworthy way. Participants also exhibited an indication that they perceived my commonality with them as an indicator that I am able to relate and

appropriately empathize with their experiences. I believe, it made participants feel understood and valued and, in turn, increased their enthusiasm to be candid in the information they provided.

The credibility of the findings is often called into question in qualitative research. Thus, one of the techniques for safeguarding against invalidity is utilizing multiple sources to uncover common ideas or themes, also known as “cross-checking” your data (O'Donoghue & Punch, 2003). To increase the confidence in the validity of this study's interview data, quantitative instruments were used to assess concepts associated with adolescent readiness to transition (the *Thalassemia Readiness Survey*). Additionally, because participants were asked to review my understanding of their ideas and experiences, this too, supports the validity of the findings.

Chapter Four

4.0 Presentation of Findings

This chapter presents findings collected from in-depth interviews conducted with adolescents with b-TM, as they shared their experiences living with and managing their condition on the cusp of transition. Five prevalent themes were identified arising from their experiences: 1) knowledge and understanding of b-TM; 2) familial support; 3) self-reflection and disclosure; 4) comradery; and 5) readiness to transition to adult care, including the impact of a *Health Transition Specialist* as a member of a comprehensive health care team. This chapter includes a description of the setting, study sample, study participants, and the findings of my research. As indicated above, all names used in this chapter are pseudonyms.

4.1 Participant Demographics

Participants of this study consisted of five individuals receiving care for b-TM at the *Hospital for Sick Children* in Toronto, Canada; four (4) females and one male (1). All lived with their parents throughout the Greater Toronto Area and commuted to the hospital for treatment and clinical follow-up. Interviews were conducted in private areas (e.g. unoccupied treatment rooms and consultation offices). All participants were between the age of 16 to 17 years old and all were enrolled in full-time secondary school studies. With respect to the type of treatment the participants were on, all participants were on both a regular transfusion regimen (the average blood transfusion schedule was four weeks for all participants) and receiving oral iron chelation therapy medication or combination therapy.

4.2 Knowledge and Understanding of b-TM

The participants are aware of their condition and expectation of, evolving health needs; and have a history of participation within the pediatric and adolescent model of care. They now will confront a new care system of health care delivery.

4.2.1 Understanding of b-TM

Four of five participants in the study agreed that they could “somewhat” describe their condition to others in the answers they provided from the *Thalassemia Readiness Survey*. However, when asked during their interview, all suggested they should know or would like to know more about their condition and that it was difficult to describe b-TM to others. Furthermore, although all participants agreed that they knew why tests outlined in the survey were conducted, they all struggled to provide an oral explanation during interviews (with two participants avoiding providing an explanation altogether). Likewise, when asked on their survey if they could identify their current medications, all participants readily stated that they knew what they were taking and why, however, during a collective review of their surveys the majority of participants were unable to describe dosage, indications, contraindications, and adverse effects.

4.2.2 Attunement with b-TM

Participants’ views on having b-TM were mainly an outcome of attempting to manage the physical symptoms of having the condition, such as “feeling tired” or “feeling drained” or “never having time off from needing to take care of my health needs.” Participants viewed the significance of their health status in the perspective of the manner in which it impacted the “normalcy” of their lives. That is, they did not focus on b-TM itself but rather their capacity to function with the condition. For example, they shared instances of “missing out” on occasions to “stay overnight with friends”; reports of being unable (e.g. feeling too tired) to participate in activities too close to their

next blood transfusion date or to “doing the things I like to do, like riding my bike or other activities,” as well as, the impact of chelation therapy on their independence (e.g. using an ambulatory pump meant staying close to home in case there were any issues). It appeared that for several participants, the daily activity of chelation therapy became an inconvenient but necessary routine, and for others, chelation therapy (oral or otherwise) was a constant reminder of the seriousness of their condition.

For three participants, having b-TM has become one aspect of who they are, not a separate entity or something that is outside of themselves, imposing on who they are. The incorporation of living with and managing b-TM allowed them to focus outward. Moreover, they acknowledged spending time and energy identifying activities or behaviours, which they felt would help provide better management of b-TM. The changes included diet modifications, listening to their bodies (e.g. “taking it easy”), and stress management. They frequently talked of “doing research” on themselves or “experimenting” with various approaches to illness management (e.g. trying yoga and meditation).

In one example, Fiona discussed how she does her best to manage both her condition and the medical care related to b-TM. She acknowledged that she might not always be able to control “*everything about it*,” but that she felt she had a role in directing its management. Fiona valued having knowledge about b-TM and believed it helped her in living with and managing it. She identified herself as very “*curious*” and felt responsible to know as much as possible about b-TM as she could, including its treatment, current research, and alternative treatment options (e.g. holistic and dietary approaches). Fiona sought information from a variety of resources. She used the internet and sought information from both her health care team at the clinic, as well as, other health professionals to learn about b-TM:

My parents have always tried to teach me to find the positive in having thalassemia and they always have explained what they could do to me I have a lot of things I want to do [in life] so I need to do the best I can to take care of my body and do all my treatments and do all my tests.

Another participant also agreed that she had a responsibility to take care of her body. Jasmine stated, "... like, we are only given this one body, so I am responsible for taking care of everything I can control, even though there may be things I cannot [control]."

4.2.3 Building a Culture of Giving Back

Several participants also spoke about the potential to integrate the knowledge gained from their experiences of living with and managing b-TM into their future professional ambitions. Flora poignantly described this connection:

I am not sure exactly what I want to do yet [pause] but I am thinking maybe nursing. ... I feel like my own experiences would make me a great nurse because I would understand what patients are going through. I know that their experiences may not be exactly the same as mine but I think, generally, people going into the hospital go through a lot of the same things.

Similarly, Jasmine, enthusiastically described her own aspirations of wanting to get into the field of medicine as a young child:

I think I always knew I wanted to be a nurse or something like that. My sister would always make fun of me for playing with the syringes and stuff [other supplies] but I was always fascinated with them and they kinda just became a normal part of my life. Now I see, that not everyone has to deal with this kinda stuff but think I might be able to help those that have too.

In all three circumstances, participants appeared to accept that their personal life experiences were "an essential resource" for being able to support others who may share the need to access health care and manage or cope with chronic illness. Participants discussed elements of their future roles as health care professionals as including: being compassionate, empathetic, and understanding, and as having credibility, and more insight into the feelings and daily experiences of those living with illness more so than other health care professionals.

Finally, several participants expressed gratitude and appreciation for the opportunity to take part in the study, "knowing that you are try to help other people understand what living with

thal[assemia] is like is so awesome ... and that according to Fiona "makes [her] happy to think it might help someone else with it [thalassemia] ...”

4.3 Familial Support

The familial relationship played a significant role in the transition to self-management for b-TM participants. It appeared that among participants of this study, families were the most knowledgeable regarding participants’ diagnosis, treatment and/or management needs, challenges and concerns/fears. Although at times some participants felt annoyed when they thought their parents were overly involved, they all unreservedly expressed appreciation and comfort in the assistance they provided. Many participants discussed how their families played a significant role in living with, and especially, managing b-TM. Moreover, among these discussions was much emotional discourse (and visual observations) about the pressures participants felt their families were put under because of their condition. Several participants became emotional during this part of the interview and clearly articulated that this topic “was hard to discuss.”

It is quite apparent that the majority of participants in this study are fairly attuned to the support that their family offer them on a daily basis. Furthermore, although some participants felt their parents were not as knowledgeable as they would like for them to be on matters surrounding b-TM, they still acknowledged the need for their support while living with and managing the condition.

4.3.1 Teaching Self-Management

Several participants discussed self-management strategies taught by their parents to promote self-management of b-TM, as well as, potential issues surrounding independence, normalcy, and self-advocacy. For example, Jasmine described the following self-management training from her mother: "... when I had my pump, she [mother] would always make me help her get out the medication and

stuff [supplies]. I think as I got older this helped me remember what to do...” She also discussed distraction strategies taught to her by her mother as an effective self-management practice “... she [mother] would make me count to 10 after my needle was put in and this helped me stay calm.”

Most participants acknowledged the efforts and commitment that their parents have put into keeping them as healthy as possible. Belle shares this sentiment, “... without my parents I wouldn’t be here, you know.” Similarly, Fiona added, “my parents never hold me back from doing anything, or treat me differently than my brothers but ... [pause] at the same time, I know they never stop looking out for my best interests either.”

Other participants shared their experience of learning to become self-advocates from their parents. When asked to provide an example of how her parents supported her, Fiona offered this explanation, “well, my parents have always encouraged me to speak up for myself, not just about thal[assemia] but about anything. Like, if my IV was hurting she would encourage me to explain to my nurse what was going on or what I was feeling.” She continued, “... I think this helped me become independent because I had to learn to speak up for myself.”

Another participant described how her family’s interaction with spirituality was one thing that helped her manage stressful periods of managing b-TM. Fiona stated:

My family is very spiritual ... this has taught me to just take each day as it comes ... having thalassemia, it really changes your perspective on life as you get older, like in terms of how you deal with things, at least for me ... my faith has gotten me through a lot of things ... [pause] and like I said, my family has always tried to think positively and [pause] I guess, I tell myself things happen for a reason

Interestingly, two participants did feel that their parents did not always understand what was going on with their thalassemia. Furthermore, they seemed to relate their parents’ lack of understanding to the struggle for them to become independent. Belle shared, “I don’t really talk to my parents about too much... I don’t think they get it and then I get frustrated, so [pause] I usually just

wait until I get to clinic to ask questions or whatever...” Similarly, Flora expressed frustration that her parents did not always have answers for her questions, “...I feel like they didn’t know a lot about thal[asemia], so I think they supported me like, emotionally and stuff, but I had to find a lot out on my own.”

4.3.2 Caregiver Burden associated with b-TM

Participants acknowledged the effort put forth by their parents/ guardians to make sure they were taken care of, in terms of their b-TM treatment and management. Many of them described scenarios involving missed work, acquiring babysitters for siblings, costs associated with travelling to and from the hospital, and extra stress placed on the household. For example, Jasmine shared:

... my mom usually comes [to the hospital] with me. But she had to work today so my sister took the day off instead. ... I feel bad about her [sister] having to come with me but I am really glad to have the company ‘cause sometimes I don’t feel that well or can’t leave the floor on my own ...

In another example,

When I was younger one of parents or my grandmother was always with me. That was a real pain for them, I’m sure ... I come to the hospital on my own now but it still costs a lot for me to get here from [city’s name] ... my mom still packs me food and stuff to bring ...

Another participant further explains,

[emotional – fighting back tears] ... like, my family does a lot for me. Like bringing me to my appointments and my mom makes sure I take my medication, like every night. And like, growing up it really sucks that they had to do so much for me ...

Finally,

[emotionally reflecting] I know me having thal[asemia] puts extra pressure on my family. Sometimes, I really get down when I think about it. Its like, for 17 years they have been doing all this stuff for me that, if I didn’t have thal[asemia] they wouldn’t have to think about – medications, iron levels, MRI’s ... I don’t know, I just feel bad about it sometimes ...

Some participants even reflected on how they perceived the onset of an unexpected diagnosis related to additional stress being placed on parental shoulders. In one instance, Jasmine explains:

I don't think my parents even knew what thal[assemia] was before I was diagnosed. I really can't even imagine what that must have been like.... They had other kids too, so it couldn't have been easy to find that out.

4.4 Self-Reflection and Disclosure

The interview data collected from participants on self-reflection and disclosure suggest that, a sense of being treated differently by others because of their condition is one of the most prominent issues surrounding living with and managing b-TM. Participants offered ideas of what “might” happen if those outside their immediate social circles were to discover their health status. Additionally, participants' experiences spoke of a sense of being vulnerable to possible harm by outside forces, most often characterized by the perceived judgments of others or by living with and managing b-TM.

4.4.1 Fear of Disclosure and Stigmatization

Fear of disclosing their health condition to others was a central theme, which emerged from this study. This theme addresses the paradox among participants of revealing versus concealing their disease condition. Generally, participants were concerned with how they may be defined or judged, if others knew of their circumstances. Each of the participants brought up the notion of disclosure being difficult. Knowing what and when to share with the various groups of individuals in their lives appeared to be an intricate consideration. Additionally, several participants appeared to worry and be concerned about if disclosing they have b-TM would be met with understanding and acceptance or rejection and ridicule. Furthermore, participants considered how disclosing that they had b-TM might impact their future aspirations.

One participant stated:

I don't really tell too many people about having thal[assemia]. Like some of my [extended] family don't even know anything is wrong. And only a few of my, like, really close friends know. And they like, know I have thal[assemia]

but don't really know what that means. I don't really want to share too much 'cause I don't want them to think I can't do things or will always be tired.

Sharing this sentiment, another participant disclosed her fear of being defined solely on the basis of having b-TM:

It's hard, you know, I don't really tell them [friends] too much. And anything they do know [about thalassemia] is something my mom has told them or their parents.... [Jasmine begins to get emotional] ... Sometimes I wish they didn't know anything though. I don't want them to think having thal[assemia] defines who I am...

Jasmine's experience provides evidence that adolescents born with chronic illness do not have a negative sense of self until they get older and are able to understand and internalize stigma.

For some participants, travelling to the hospital to receive blood transfusions and clinical appointments meant having to explain absences away from school to classmates and teachers who may not have heard of b-TM. As Fiona explained: "... I disappear from school once in a while and sometimes you know I'm at the hospital and they'd be like where did you go?" Flora shared how difficult it was to explain b-TM "I wish I knew how to explain it to them without freaking them [friends] out." Others expressed concern of how living with and managing b-TM will impact their future ambitions and if they will be able to accomplish them at all.

I think mainly, I wonder how, [pause] I guess [pause] now that I am starting university... how... like what if I fall behind on stuff and maybe I won't always be able to be at school [pause] how I will explain why I need extra time or help...

In another example Duke explained,

I'm kind of worried about my health because I don't know how long it will be okay for in the future. Right now, I'm okay and everything, but I'm not sure when it may get worse, or if it will just stay the same....

Similarly, Belle shared,

I'm not really worried right now at 17 years old, you know, right now this isn't that bad. I can guess that at some point, in all likelihood, my thal[assemia] is going to get harder to manage and I am sure then will get a bit scarier.

Another participant questioned whether there was a need to inform her university at all, in fear that she may face the stigmatization associated with being enrolled with as a student with a “disability” stating:

I am starting to apply for university and stuff but I am wondering if its something I need to tell them. Because I don't want them to think I need any special help or anything.

One participant revealed that although her future as a person with b-TM did not cause her to worry, fear of having to disclose having the condition to employers did appear to be a potential challenge:

I am not really worried about the future. But I know I will have new challenges. Like if I get a job and how I will tell my boss about having thalassemia.

4.4.2 Overcoming Vulnerability

Participants spoke of ‘being normal’ in their life with b-TM, which often involved an altered view of *self* in which vulnerability and dependency on others was highlighted. For example, Jasmine spoke about her sense of self in relation to control over her life and the dependency she has on family to help her:

... you don't always have control and that's hard... that you have no control over the scheduling of the appointments or tests you need, no control if you have a reaction or something ... I kinda have to give up control sometimes too [pause] because I needed my family to help me, like with getting me here [hospital] and reminding me about my medications and stuff ...

Other participants also shed light on their sense of vulnerability, which stemmed from the constant reminder of having b-TM through the experience of physical symptoms. For example, Duke stated, “I can tell how close I am to a [blood] transfusion date based on how tired I am. The more tired I am, closer I am to my next [blood] transfusion.” Another participant, Fiona, acknowledged how scheduling her blood transfusion dates is closely related to physical symptoms stating:

I try to come [to clinic] on Friday's so I can rest over the weekend. 'Cause sometimes coming to the hospital for my [blood] transfusions makes me really exhausted. But this way, I can rest and start my week feeling full of energy and refreshed.

The same participant discussed the challenges of having to depend on the health care system while fighting to gain independence in almost every other aspect of her life. For example, Fiona states:

It's impossible not to have to depend on someone or the hospital for some things. Like when I had my PORT [port-a-cath or a central venous catheter] I had to come to the hospital all the time ... that's kinda hard 'cause on one hand, I just want to live my life and like, go to homework club but there is a constant reminder of things I know I have to take care of first.

Interestingly, none of the participants of this study described any specific experiences in which their perceived fears of rejection, pity, and isolation from their social interactions actually materialized.

4.5 Connecting through Comradery

In the notion of having a sense of mutual understanding/ community between two people or a group of people sharing the same circumstances, comradery was, surprisingly, an avenue in creating a novel method in gaining understanding of my participants' experiences. All of the participants in this study appeared to have a positive reaction when they learned I too, was a thalassemia patient. One participant (Flora) expressed her surprise and admiration, "Wow. You're really a patient too? Here [pause] in Toronto? ... That's amazing. I would have never thought you were a patient." Similarly, another participant (Fiona) was taken back by the possibility that someone who had b-TM had the potential to enroll in graduate studies and further to professionally contribute to the field of medicine, stating, "That's the coolest thing. That you're a patient and in graduate school. Do you know if there are any people with thalassemia who are doctors or nurses?"

The implied sense that WE have shared experiences (or commonalities) seemed to create an

instantaneous connection, a direct understanding of how OUR lives are shaped by living with, and managing b-TM. Although I did not disclose too much of my own personal struggles (I only stated that there were/ are many parallels with my own experiences), it appeared that the experiences of my participants were enough to appreciate and understand what my OWN life must be like living with b-TM. For example, *“That’s crazy that you are a patient too. Ha ha ... so you “totally get everything I said ha ha...”*, or *“That’s really awesome. Would you mind if I asked you some questions too? Like about school and stuff...”* Several participants took the opportunity to ask me their own questions, ranging from university experiences to my questions pertaining to my own health status (e.g. chelation medication experiences, current iron burden levels, etc.).

The conversations after my own health status was disclosed to participants was particularly eye-opening because it demonstrates that although the majority of participants reported that they had a good relationship with their health care team and knew where to seek out information pertaining to b-TM, there may be a benefit for those with the condition to have an opportunity to engage in and intermingle with older “adults” that have similar experiences living with and managing b-TM.

4.6 Readiness to Transition

Participants’ general attitude toward transition was found to be moderately positive. For example, although Duke “... didn't see pediatric and adult care different... just another hospital to get treatment at," the majority of participants indicated they had some apprehension about certain aspects (new environment, building new relationships with staff, uncertainty of new facility, getting used to a new system, etc.) and admitted to being concerned about transition to a little or even a large extent. Participants described a close emotional attachment to their current pediatric clinic and current health care team. Additionally, participants expressed an ambiguous awareness of how their treatment

routine may change, which was mostly founded on hearsay rather than tangible information (e.g. through older siblings or members of their current health care team).

Participants desired to attain more knowledge on the transfer process, to acquire strategies to get to know their new health care team and to have enough time to adjust to a new system or “system change. Overall, all participants appeared to have a positive outlook on transitioning from pediatric to adult institutions of care in the near future. For example, Belle shared, “I am kinda sad about having to leave Sick Kids... [pause]... It’s kinda like a second home for me but I know it’s a normal part of growing up. And I am excited for what that means too.”

4.6.1 Self-transcending into Adulthood

The transfer from pediatric to adult care, was explained by all participants as a symbolic entry into adulthood; a groundbreaking self-management achievement, representing a change in self-identity described as, "... growing-up ... getting older ... not being a kid any longer," ... or as a time when you are prepared "... to go out into the real world and make choices as an adult patient."

I think it will be fine. I know I am going to have to step-up and take care of myself more ... I will have to make my own choices and learn a lot more but I have an older brother who can help me because he has thal[assemia] too and that’s kinda what has happened my whole life. ... I learn from him or he tells me new things.

Similarly,

I don’t think it will be too different [transitioning to an adult hospital]. It will mean I make my own choices. ... I already come to the hospital on my own now so I really don’t think it will be that different , like not a huge deal or anything ... But I hope we aren’t just rushed there and we get to like meet everyone and learn about the [new] system.

Another participant shared her apprehensions about transitioning to adult orientated health care:

I know I will eventually have to leave Sick Kids ... and not being familiar with the new hospital and all the in’s and out’s. That's where the anxiety comes into play, I guess ha ha. I'm all about ... I guess I am not very good with change

All participants agreed that 18 years of age was an appropriate age to transition to an adult hospital. However, there were various reasons given for this sentiment. Flora explained, I think 18 is a good age to transition because it's a natural point in a person's life where they are making independent decisions anyways. However, some participants felt that chronological age was not an important indicator on transitional readiness as maturity level and capacity to self-manage their condition. For example, Jasmine states:

I think age has a lot to do with it but also if a patient isn't comfortable they shouldn't be forced to transition to another hospital. It's probably better to decide if a person should transfer based on if they are, like mentally, really ready or not.

4.6.2 Therapeutic Relationship with the Health Care Team

Regardless of the perceived level of preparation, all participants described the thought of transitioning to a new hospital as "a bit scary... at first" and placed much of their presumed future success with the notion that they had a positive relationship with their current health care team. As Fiona explained:

... you know in life, with anything change brings anxiety, and sometimes you might be afraid because you do not know what to expect because you are used to your care being a certain way, so now when you're moved to a different or new hospital you don't know the new people who will care for you. You haven't developed a relationship with them, so you really don't know what you're gonna get yourself into.

Several also acknowledged the idea of "being spoiled" or "not having to worry about too much here" (as far as appointments and understanding test results) at the *Hospital for Sick Children*. For example, Belle discussed not having to schedule any appointments herself and having to just "show-up" when she was told. She explained:

I think I know what tests should be done. And if something doesn't get booked in like, years – like my hearing test, I would say something but really there isn't too much I do, in terms of booking my own tests or understanding results. My [health care team] always checks in with me and is really good at keeping me informed. I

guess, we are kind of spoiled here, ha ha.

Participant interviews highlighted the value placed on health care team relationships, provider expertise and being included in conversations and decision-making surrounding their care. Every participant expressed these ideas several times throughout their interviews. Several also discussed experiencing feelings of loss when relationships are terminated (e.g. exceptional nurses leaving) and confirmed their appreciation when they felt their health care teams acknowledging their aptitude in making decisions.

4.6.3 Gaining Independence

Participants discussed "taking control" of their management as a gradual process of "...learning when I am supposed to do when I was little and eventually ... now I do it without thinking ... being more responsible for my own care ..." Most appeared to realize that the role their families played in managing b-TM might change and perhaps, "...my mother and father won't always be there to hold my hand and tell me what to do or how to do it; I would have to learn to do things on my own..." and the need to "really step-up." As the interview progressed, it appeared several participants really started to reflect on how their self-management had evolved since their childhood.

Belle recalled:

I guess, when I think about it having thal[asemia] made me independent very early. Like, I can remember being 4 or 5 years old and wearing my pump – I was in charge of getting together most of the supplies and --- if my needle fell out while I was sleeping I was responsible to go and wake my mother or grandmother to restart the whole thing. So, I guess that was one way I was kind of being taught to take responsibility for myself.

Similarly, Jasmine discussed her evolution to self-management as a consequence of her desire to be independent. She explains:

Taking the pills now instead of a pump is like a miracle. I can take them whenever I want and if I am going out I can just bring them with me. I know how important it is to take my medication. I don't want to start having any problems

[with iron overload]. So I guess, I started like really paying attention [to self-management] as soon as I became a teenager... I didn't want to be in the hospital. I wanted to be out with my friends, having fun and I knew if I got real sick I wouldn't be able to do those things.

Although several participants confessed, "I still wouldn't be able to do without my family," the majority of the group reported needing only "once in awhile" reminders regarding taking medications and attending appointments" or not "overdoing myself." Two participants did acknowledge that sometimes the advice they received from their parents would go ignored, "I just do my own thing because I don't really think my parents get it," but for most, advice was sought only "... if I totally don't know what to do; but most of the time I know what I have to do better than my [parent]."

4.6.4 Health Transition Specialist Role in Preparing for Transfer to Adult Orientated Care

Innovatively, the *Hospital for Sick Children* integrated a specialized role for a *Health Transition Specialist* within the hemoglobinopathy clinic, whose role includes supporting adolescents and young adults with red blood cell disorders by aiding in the preparation for and throughout the transition from pediatric to adult health care. According to the hemoglobinopathy team, this role was implemented within the clinic mainly to assess adolescents' and young adults' readiness for transfer and knowledge of health condition at multiple time points; to provide tailored education, guidance and support to adolescents' in developing self-management skills; and to facilitate smooth transition between pediatric and the adult institution of care.

Although, the implications of this role have not been critically evaluated, every participant appeared to have been positively impacted by having a *Health Transition Specialist* as a regular member of their health care team. Fiona stated, "I know I would have way more anxiety and worries and be stressed out, if I didn't have [transitional coordinator]." Furthermore, several participants described feeling "unprepared" for transfer before meeting and working with the clinic's new *Health*

Transition Specialist:

Well, I think I just kept telling myself it [transferring to an adult hospital] wasn't going to happen soon ha ha. I think I was like 15 when I was told "you'd better start thinking about what happens when an adult, like 18 or whatever" and I always kept pushing that in back of my mind. Now, I've gotten to talk with [transitional coordinator] and I guess like its so much more real. I feel like talking with [transitional coordinator] has been extremely helpful about moving to a new hospital.

Another participant explains,

Yeah, [transitional coordinator] is awesome. Like they [health care team] are always encouraging me and telling me there isn't anything I can't do, but you know, [pause]... they probably have to tell us that ha ha. So it means a lot to be able to have [transitional coordinator]...

Overall, the benefits of including a *Health Transition Specialist* as an endorsed clinical team member appears promising for improving the outlook among vulnerable adolescents with b-TM.

Chapter Five

5.0 Discussion of Findings

The previous chapter identified five broad qualitative themes: knowledge and understanding of b-TM; familial support; self-reflection and disclosure; comradery, and readiness to transition, including the impact of having a *Health Transition Specialist* as a member of a comprehensive health care team. In this chapter these themes are discussed in relation of how they tie into three comprehensive topics; the development of self-efficacy in adolescents living with, and managing b-TM; fostering self-management practices; and adolescents' attitudes and concerns regarding the transfer process from pediatric to adult orientated institutions of care. Additionally, clinical implications resulting from this research study, the study's strengths and limitations, my personal research reflections, as well as, concluding thoughts are presented.

5.1 Development of Self-Efficacy in Adolescents Living with, and Managing b-TM

The transition to adult orientated health care institutions for individuals with b-TM represents a significant milestone in the life of an adolescent. However, with this milestone come new responsibilities, including: medical decision making, autonomous self-management practices, and advocating on their own behalf. Self-efficacy can be defined as an individual's capacity to attempt difficult or novel tasks, and/ or to cope with adversity arising from complex or demanding circumstances (Cross, March, Lapsley, Byrne & Brooks, 2005; Luszczynska, Scholz, & Schwarzer, 2005). In a study evaluating the indicators with the greatest impact on successful transition for adolescents diagnosed with *Sickle Cell Disease*, Sobota, Shah and Mack (2016) identified self-efficacy in disease management as one of the highest predicting factors of successful transition. Self-efficacy has also been included in the evaluations of transition readiness in various studies on the

transition process of adolescents with chronic conditions (Treadwell et al., 2016; Sobota, et al., 2014; Sawicki et al., 2009), which demonstrated that increased self-efficacy was associated with improved transition to adult orientated health care institutions (Molter & Abrahamson, 2015). Furthermore, other research has provided evidence to suggest that self-efficacy can be used as a predictor of positive health outcomes such as, adherence (taking medication), health behaviours (eating right or engaging in physical activities), and overall chronic illness management (Bar-Mor, Bar-Tal, Krulik & Zeevi, 2000; van Es et al., 2002; Griva, Myers, & Newman, 2000; Ott et al., 2000). Additionally, self-efficacy may also impact adolescents' physical, emotional, and social quality of life (Griva, Myers, & Newman, 2000; DiNapoli & Murphy, 2002). Similarly, van Schayck et al., (2007) credit the successes of self-management on clinical and functional outcomes as a direct result of formal education initiatives intended to construct knowledge, improve overall quality of life satisfaction, and strengthen self-efficacy.

In the current study, participants relied to a great extent on their parents (or guardians) and health care team for health care decision-making and the management of their condition (e.g. scheduling tests, following up on results, medication reminders, and understanding symptoms). Although in their survey responses most participants reported an understanding of managing b-TM, the majority were unable or hesitant to clearly describe “what tests” should be completed and when, how to interpret test results; the purpose of the medications they were taking (e.g. describing chelation medication as “iron pills” instead of “iron removing” medication), and also admitted to a consistent need to be reminded to take medications and/ or complete specific clinical tests. Overall participants lacked insight into the complexities of attaining an optimal level of health. As such, the findings suggest the importance of interventions that target an increase in self-efficacy among adolescents with b-TM, specifically focusing on those that aim to promote confidence and the ability

to effectively take on more health related responsibilities pertaining to living with and managing b-TM.

5.1.2 Facilitating the Empowerment of Adolescents with b-TM

A consistent definition of patient empowerment is difficult to find in healthcare literature, however most descriptions relate the concept to self-determining individuals that have some control over their own health and healthcare decision making, rather than being passive receivers of their healthcare (Aujoulat, d'Hoore & Deccache, 2007; Aujoulat, Marcolongo, Bonadiman & Deccache, 2008; Funnell & Anderson, 2003; Lorig & Holman, 2003). Additionally, related to this notion, are opinions of patient compliance and adherence. Compliance refers to patients surrendering to the authority of healthcare providers, whereas, adherence refers to patients voluntarily agreeing with their healthcare providers, maybe in part due to mutually agreed upon shared-decision making (MacStravic, 2005).

Researchers believe that facilitating patient empowerment will enable those with chronic healthcare needs to, 1) make more rationale healthcare decisions, maximizing their health and overall wellbeing; 2) decrease a dependence on healthcare services; and 3) contribute to a more efficient use of healthcare resources (Aujoulat, Marcolongo, Bonadiman & Deccache, 2008; Funnell & Anderson, 2003; Lorig & Holman, 2003). It is important to understand that culture, age, and socio-economics would unquestionably effect empowerment and the degree to which different populations or groups can be, and wish to be, empowered. For individuals with complex healthcare needs (e.g., adolescents learning to live with and manage b-TM), empowerment can be facilitated not only by healthcare professionals but by their parents or guardians through various modes of education (e.g. self-education facilitated by the internet; specialized educational programs or initiatives; peer-mentoring program, etc.), counseling, and encouragement to take on more age-appropriate health-related

responsibilities (Coulter, 1999; Holmström & Röing, 2010; Bravo et. al, 2015; Ho, Jameson, & Eiser, 2017). Empowered adolescents with b-TM will be equipped to interact with an adult-orientated healthcare system, and will be independent and actively involved in the decision making process regarding their healthcare.

5.2 Fostering Self-Management Practices

Adolescents transitioning from pediatric to adult institutions of care are expected to be autonomous and assume an active role in their health care, however this is sometimes a challenge that they are not sufficiently prepared to meet (Ostbye, 2005; Huang et al., 2011). Developing effective self-management practices ahead of transfer to adult institutions will better prepare adolescents and their families to adjust to a new health care environment. Given the extensive role that parents (and/or guardians) (Powers & Dell-Orto, 2004), as well as health care professionals (Dwyer-Matzky, Blatt, Asselin & Wood, 2018), are perceived to play in adolescents' development of self-management practices, age-appropriate gradual shifts in responsibility for b-TM management should be strongly considered.

5.2.1 Role of Family

Not surprisingly, families play a significant role in how individuals cope and react to living with a chronic illness (Powers & Dell-Orto, 2004). The data I collected regarding the familial relationships of my participants was detailed and abundant. Not only did participants openly discuss the relationships they had with their parents (or guardians), but also several of them opted to discuss the relationships between their siblings and other extended family members. While participants had a variety of experiences of how their families support them in living with and managing b-TM, it was quite clear that their families played a central role in, their understanding and knowledge of b-TM;

how they coped with the challenges of living with their chronic illness (including the attainment of self-management practices); and, how their families provided a means of different levels of support (e.g. emotional, financial, etc.). Several of the participants were emotional when they shared experiences about familial support. Furthermore, most of them appeared to fully appreciate the time and efforts of their families in doing their best to keep them as healthy as possible, even though this left some of them feeling like a burden at times.

The participants described familial behaviours that reflected their parents' (or guardians) emphasis on normalcy and being vulnerable. Participants in this study shared their experiences on how their families interacted with them (and taught them), provided encouragement, and/ or promoted normalcy. However, some participants also provided explanations or insight on issues of "privacy" and "independence" in their families and the struggle between parents and themselves. In one situation, a participant perceived that her parents overstepped boundaries often and didn't understand her condition or circumstances. Scenarios, such as the one described above, may impact the possibility of conflict within a family's dynamic (Wong et al., 2010). Furthermore, parents may have difficulty stepping back from the roles of "health care manager" when appropriate. In a longitudinal study conducted by Lorig et al. (2001), the implementation of a low-cost program for promoting age appropriate health self-management practices among parents and their adolescent children found a significant improvement in overall health status and satisfaction, while also reducing health care costs associated with populations of various chronic diseases.

These interpretations may suggest that for individuals with b-TM, familial support plays a particularly protective role with respect to the teaching of self-management practices. Past educational research has demonstrated that familial involvement is a contributing factor to successful

transitions, especially for young adults with chronic illness or disabilities (Greene, 1996; Halpern, Yovanoff, Doren, & Benz, 1995).

5.2.2 Health care Providers as Enablers and Collaborators

The patient and health care provider relationship is considered a fundamental element in the delivery and success of health care programs (Bowen, Henske, & Potter, 2010). Through the review of participant transcripts, it appeared that there existed what could be labelled as an “us versus them” mentality, with the belief that there is a distinct difference and separation between the members of a health care team and those they treat. When asked about how their health care team supported them, participants spoke positively of their person-to-person interactions with members of their health care teams, but for the most part, expressed a sense of apprehension when referring to the health care delivery system as a whole. This may have been a result of participants fearing that, I too, was a strict member of a research team assigned by their governing health care institution. However, I am more inclined to believe that for most individuals with b-TM, engagement in the health care system normally involves trying to negotiate appropriate chronic care in an acute health care system, a system designed to cure or control symptoms. This acute bio-medical health care model is one that focuses on the physiological aspects of diseases, but under appreciates the psychosocial experiences and/ or meanings that are a part of managing chronic illness. As this study illustrates, and as several other studies have demonstrated before it, psychosocial concerns (e.g. self-reflection, need of various support systems, fear of disclosure, etc.) are a substantial cause of the burdensome nature of living with a chronic illness (Gysels et al., 2007; Hernandez, Balter, Bourbeau, & Hodder, 2009; Ironside et al., 2003; Koch, Jenkin, & Kralik, 2004). Since the present bio-medical models of care neglects these types of concerns, it hinders health care team professionals from most effectively addressing the needs of particular populations of vulnerable patients (Kleinman, 1988, p. 9).

Identifying the manner in which adolescents perceive their support systems provides important information for planning interventions and educational initiatives that could reduce the prevalence of non-adherence to chelation therapy among this population. For instance, given the insight provided by participants, it may be more effective and appropriate to develop familial education programs that aid parents (or guardians) in age-appropriate tools to teach self-management practices for developing children and adolescents with b-TM. Additionally, fostering and developing strong collaborative relationships between adolescents and their health care teams and health care delivery systems may provide a fundamental opportunity to engage in meaningful conversations about adherence and proactive management of b-TM (one means in which to facilitate these relationships may be through the use of a *Health Transition Specialist*). Moreover, it may establish a trusted connection between adolescents seeking information pertaining to b-TM and those considered knowledgeable on the condition.

5.3 Adolescents' Attitudes and Concerns Regarding the Transfer Process

Most participants' expressed a positive outlook on their anticipated transfer to an adult orientated health care institution. However, many of the same participants also expressed and shared their anxieties and apprehensions about leaving their current health care providers. As such, at the time of their interviews, it appeared that participants felt unprepared to transfer. Perceived challenges of transitioning to an adult health care institution included: having to "start from the beginning with a new health care team," having to navigate through a new environment, and generally, fear about the unknown. The findings of this study are similar to those discussed elsewhere: "transitioning into an unfamiliar adult environment was instrumental in promoting anxiety and fear of the unknown" (Bryant, Young, Cesario, & Binder, 2011: 278).

For participants living with and managing b-TM, the beliefs and perceptions about the adult-oriented health care systems appeared to increase their anxieties and pose unanticipated questions of their future care. Tuchman, Slap and Britto (2008) and Houtrow and Newacheck (2008), examined the experiences and expectations of adolescents with chronic health needs who were apprehensive of the transition to adult orientated institutions of care. From their research the following recommendations were provided: an early awareness of the planned transition, an orientation and introduction to the new institution of care, and a structured transition program to assist with transfer (Tuchman, Slap and Britto, 2008; Houtrow & Newacheck, 2008). The incorporation of these recommendations may effectively alleviate the worries that adolescents with b-TM have regarding transition from pediatric to adult health care institutions. Furthermore, the health care transition for adolescents with b-TM should be a gradual progression, designed to meet the specific needs and achieved with an appreciation of the developmental and psychosocial transitions simultaneously taking place.

5.4 Clinical Implications Stemming from this Research Study

Three central clinical implications stem from this research: 1) the development of age-appropriate educational resources for developing individuals with b-TM; 2) an opportunity for a peer-mentoring should be considered; and 3) the role and implementation of a *Health Transition Specialist* into the current comprehensive health care team should be critically evaluated.

5.4.1 Development of Educational Resources

As adolescents take on a more active role in their health care decisions, it becomes necessary to develop and provide additional educational resources aside from a physician's knowledge (Bourgeois, et al., 2008). Research is needed that investigates factors that influence self-management

and identifies effective strategies to aid adolescents with b-TM during their transition from pediatric to adult orientated care (Nguyen et al., 2016; Modi et al., 2012). In doing so, health care professionals will gain insight into the development of self-management practices, identify those at risk for poor transition, and improve or develop new patient-orientated tools (Modi et al., 2012). This insight will also help health care professionals to aid adolescents with b-TM as they attempt to gain more independence and confidence in their ability to successfully live with, and manage their condition.

As two participants pointed out, current information communication technology (ICT) provides an avenue to educate and support adolescents with chronic illnesses. Additionally, ICT provides a source of support for those with chronic illness by offering resources outside of the traditional hospital or health care environment (Bourgeois, et al., 2008). It also provides a platform for easier accessibility and patient-orientated interaction. For example, in a study investigating the advantages of an *Electronic Support Group*, researchers found that adolescent participants with cystic fibrosis acquired knowledge, self-esteem, and a sense of community as they communicated with others living with the same condition (Johnson, Ravert, & Everton, 2001). Similarly, in another study conducted on the use of an electronic *Self-Directed Individualized Education Program* among youths with a range of disabilities, Seong, Wehmeyer, Palmer and Little (2014) found an increase in level of self-determination in participants, and a positive impact on transition knowledge when compared to a placebo-control group. As several of the participants of this study admitted to using ICT as a resource to seek out answers to their health related questions, there may be a significant benefit in developing effective and reliable patient-orientated ICT educational tools for adolescents with b-TM.

5.4.2 Consideration of a Peer-Mentoring Initiative

Mentoring is considered a process by which, “an individual with more lived experience provides knowledge and insight to an individual with less experience” (Peyton, Morton, Perkins, &

Dougherty, 2001, p.351). Current literature on peer-mentoring programs suggests that there may be significant benefits to both mentors and mentees (Ahola Kohut, Stinson, Forgeron, Luca & Harris, 2017) and, that these programs may facilitate smoother transition periods during participants' lives (Carragher & McGaughey, 2016). Furthermore, several studies have examined the impact of mentoring programs on adolescents living with a chronic condition and found a positive association between mentoring and improved mood, attitude towards self-management, and self-esteem (DeWit, DuBois, Erdem, Larose & Lipman, 2016; Merianos, King, Vidourek & Nabors, 2015; Maslow & Chung, 2013; Herrera, DuBois, Grossman, 2013; DuBois, Portillo, Rhodes, Silverthorn & Valentine, 2011). Mentors, on the other hand, appear to benefit from their role in the form of new informational support (e.g. new coping strategies), emotional support from both those they are mentoring and other mentors (Mackner, Ruff & Vannatta, 2014), as well as, an enhanced self-evaluation (despite no change in personal circumstance) (Lipman, DeWit, DuBois, Larose & Erdem, 2018).

Other studies investigated the impact of peer-mentoring initiatives on predictors of medical transition for those with complex chronic illnesses; self-management practices, knowledge, and attitude on transiting to adult care (Jerson et al., 2013; Rhee, Belyea, Hunt & Brasch, 2011; O'Mahar, Holmbeck, Jandasek & Zukerman, 2009). The studies concluded that the utilization of peer-mentors was a safe, practical, and effective option to improve adherence, knowledge, and self-management practices of adolescents' anticipating and/ or during transfer to adult care (Jerson et al., 2013; Rhee, Belyea, Hunt & Brasch, 2011; O'Mahar, Holmbeck, Jandasek & Zukerman, 2009).

Taken together the research suggests that the peer-mentorship programs may be beneficial to both mentees and mentors. Becoming a mentor allows those with real, lived experience to share their "wisdom" with less experienced individuals and may also develop a strong connection to their identity as someone living with b-TM. Additionally, as there appeared to be some positive outcomes

regarding improved medication adherence among adolescents who had a peer-mentor, there appears to be a significant opportunity in the potential of this approach. A peer-mentoring program will allow adolescents with b-TM the opportunity to make sense of their experiences within the context of the lived experiences of their peers.

Moreover, as mentors reflect on their own experiences and the lessons they have learned, those they are mentoring may gain invaluable insight not considered or appreciated by their health care teams. For example, although all of the participants of this study concluded that oral chelating medications were “easier” to administer than that of the ones received via an ambulatory subcutaneous infusion, they also brought forth unique challenges and participants often felt that their health care team failed to recognize this. Given the surprising finding of this research (re: the information acquired after my own health status disclosure), there exists evidence to suggest that adolescents living with, and managing b-TM may benefit from a peer-mentor in supporting their anticipated transition to adult care.

5.4.3 Implementation of a Structured Transition Plan, including the addition of a Health Transition Specialist into Clinical Management

Current literature has recognized the reluctance of some adolescents with chronic illnesses to leave the care of pediatric hospitals, and has identified feelings of anxiousness, worry, and fear (Boyle et al., 2001; McCurdy et al., 2006). Additionally, this literature provides consistent evidence that most youths and adolescents, including those with complex health care needs and their parents receive limited or ineffective transition planning (Blinder et al, 2013; Garvey et al., 2013; McManus et al., 2013; Kakkar et al., 2016; Knight et al., 2016; Kuhlthau et al., 2016), including the most suitable time or age to transition to adult care (Dobson, Bryce, Glaeser, & Losek, 2007). Together

these challenges contribute to preventable health related consequences and overall poor health outcomes.

With the growing recognition of the challenges adolescents' with complex health needs face, as well as, acknowledging the greater demands placed on adult specialists to treat these adolescents, in January 2012, the *Canadian Association of Paediatric Health Centres* (CAPHC) established 19 recommendations to enhance and guide the care of youths with special health care needs through adolescence into adulthood. The objective of these guidelines include: "1) to influence transitioning at the person and clinical level, prompting change over time to the system level; 2) to provide a framework for a supportive process for transitioning from pediatric to adult health services; and 3) to identify collaborative processes, tools and resources for all stakeholders in the transition of youth to adult healthcare" (Watkins, 2016). Given the knowledge of the high stake demands that living with, and managing b-TM entails, adolescents with the condition may greatly benefit from a structured transition plan tailored to meet their specific needs. As such, developmentally appropriate, purposeful discussions involving adolescents, with and without parents, may help those with b-TM acquire the beliefs and an understanding of the need for ongoing care, including adherence to chelation therapy. Furthermore, an enhanced continuity of pediatric care and the establishment of explicit details for adult care transfer might be sufficient to improve the likelihood of a positive transfer.

Furthermore, although several studies identified the necessity for member of an adolescent's health care team to aid in the transfer process (McDonaugh, 2005), none provided a clear description of a) the member's roles and responsibilities, and b) status as belonging to a recognized health care profession (e.g. transitional navigator, coordinator, or specialist) (Crowley, Wolfe, Lock, & McKee,

2011; Grant, & Pan, 2011; Lugasi, Achille, & Stevenson, 2011; Van Walleggem, MacDonald, & Dean, 2008; McDonagh, 2005; Baldassano et al, 2002).

According to the hemoglobinopathy team I worked with, the role of the current *Health Transition Specialist* was created to: assess adolescents' and young adults' readiness for transfer and knowledge of their health condition; to provide tailored education, guidance and support to adolescents' in developing self-management skills; and, to facilitate smooth transition between pediatric and the adult institution of care that adolescent may transfer to. Additionally, the *Health Transition Specialist* is responsible for continuing to follow adolescents post-transfer in order to provide system navigation, orientation to a new environment, and to offer continued support.

Although, no study to date has evaluated the impact or effectiveness of this role, it appeared that all of the participants in this study were positively impacted by the inclusion of a *Health Transition Specialist* as a member of their current health care team. Most participants stated that they felt less apprehensive or worried about their transition because of this resource, and, at the most basic level – all participants had an opportunity to discuss the general transition process and timeline with their *Health Transition Specialist*. Although these participants may not have any experience to compare having a *Health Transition Specialist* to, the majority of them discussed their appreciation of the role and current specialist with pronounced enthusiasm. Merely taking the time to discuss the expectations and timeline of anticipated transfer may alleviate the worries that adolescents with chronic illness have towards transferring to adult orientated health care institutions (Tuchman, Slap & Britto, 2008). Given that the role of the current specialist is to introduce the concept of transfer, answer questions pertaining to transfer, and identify those at risk of poor transition it would appear that her role is fundamental in designing a successful transitional program for adolescents with b-TM.

5.5 Strengths and Limitations

The in-depth, semi-structured interviews generated detailed descriptions of the experiences of adolescent participants' on the cusp of the transition from pediatric to adult institutions of care. According to interpretive phenomenology, there are countless ways to experience and interpret lived experiences (Creswell, 2007). Adequate sample size in qualitative research continues to be a significant issue (Morse, 2007). Sample size in qualitative research is not based on attempts to generalize (Creswell, 2007), but rather to elucidate the phenomenon of interest -- in this case, transition as experienced by adolescents with b-TM. As such, a homogenous sample was purposively chosen to elicit detailed and rich accounts of the experiences of adolescents living with and managing b-TM. These experiences may be transferable to other individuals experiencing similar life circumstances of living with chronic illness. That is, although the majority of participants' experiences were related to b-TM topics (e.g., chelation therapy), there were numerous considerations that apply to other conditions – e.g., the necessity for guidance in negotiating systems of care; the fears of disclosure and stigmatization of having a chronic illness; and the necessity for fostering and developing differentiating support systems for adolescents with chronic health needs. These concerns may generally apply to adolescents' anticipating transitioning to an adult orientated health care institution regardless of underlying disease burden.

My participants' stories and experiences allowed me an opportunity to identify five reoccurring topics during my thematic analysis. In addition, the interpretative analytic approach that I adopted permitted me to gain deeper insights as to how adolescents experience living with, and managing b-TM. I was mindful of my assumptions and beliefs throughout the entire research process, keeping reflexive field notes, discussing emerging ideas and themes with my thesis committee members and research team at the *Hospital for Sick Children*, and interpreted the transcripts open-

mindedly. It was also imperative that I recognize and acknowledge that my own experiences as an individual living with b-TM, may not be able to be entirely removed from the narrative. Through this reflexivity, my readers can better appreciate the interpretations that I have made.

The aim of qualitative analysis is to obtain a comprehensive, detailed description of a phenomenon (Houghton, Casey, Shaw, & Murphy, 2013). However, similar to other types of research, the method does place constraints within a study. For example, consideration of disclosure provided by my participants. Despite ensured confidentiality, participants in this study may have been reluctant to share information that would cast negative light on their circumstances as individuals living with, and managing b-TM. This reluctance may stem from an unwillingness to share personal (and sometime difficult) experiences with a researcher they had just met. Additionally, other factors that contribute to creating an individuals identity, including the recognized influences that pertain to social determinants of health, may have influenced the information participants were willing to share during their interview, for example these may include ethnicity/ culture, family dynamic, the socioeconomic status, and/or cognitive capacity. It is important that future research in the area of b-TM consider how these factors may shape the beliefs and perceptions (and thus, experiences) of adolescent individuals with chronic health care needs, including living with, and managing b-TM.

5.6 Researcher's Reflection

Prior to this research study, my research experiences, which included collecting data on b-TM, in addition to my personal experiences living with, and learning to manage the condition, allowed me to realize that my own beliefs and perceptions, as long as they were unresolved, could significantly intrude within my interpretations. And this was further intensified by the notion, that what I chose to research was unavoidably something I feel strongly about but which also continues to

present me with new challenges regularly. While this research study surpassed my own expectations and was extremely fulfilling, it posed certain questions pertaining to my own critical reflection.

Reflectivity involves the constant shift between being part of a phenomena and being able to step back and think about it methodically, defined as "... the process of stepping back from an experience, to ponder, carefully and persistently, its meaning to the self through the development of inferences' (Daudelin, 1996: 39). Specifically, reflectivity involves the consideration of how our own values, beliefs, and experiences impact the research conclusions we have established. The ability to critically reflect is held as a significant element within healthcare research (Johns, 2004).

Additionally, critical reflection within qualitative research offers a method for gaining insight into our own professionalism. Several of my participant's stories paralleled my own experiences so closely, that at times I felt myself on the brink of an emotional response. At the conclusion of one of my interviews, and after disclosing my own health status one of my participants observed this. Jasmine informed me that she did notice that I seemed to get slightly emotional while she discussed some of her stories and wondered if I was relating with her experiences but did not want to disrupt the interview to ask. As a researcher utilizing interpretive phenomenology, I acknowledge that although two individuals may share the same diagnosis, the manner by which they experience that diagnosis will differ; even though some occurrences may be common among them.

Another challenge that I encountered was the ability to sustain my role as researcher instead of a mentor. I remember that during Belle's interview, as she discussed contemplating disclosing her health status on university applications and the inner turmoil she was going through, all I wanted to do was to give her praise and encouragement and let her know that by recognizing and appreciating the unpredictability of her health circumstances she was paving a road to success. I was surprised at my own ability to keep my "researcher hat" on and stay focused on the purpose of my interview.

Although, I do remember feeling incredibly relieved at the completion of our interview when she requested if she could “pick my brain” regarding my own university experiences. In reflecting on my field notes and Belle’s transcript, it was often difficult to see evidence of my “footprint” in the first half of her interview. I believe this is due to my strong attempt to keep a minimal presence prior to disclosing my own health status. This also allowed me to think introspectively, as I contemplated how having b-TM impacted adolescents as they thought of their futures beyond the scope of their health status.

The connection that I developed with these five participants was extremely rewarding and I feel so incredibly honoured to have had the privilege of listening to their personal journey’s living with, and managing b-TM. The strength and courage of these participants, on top of the rich data I was able to collect, has inspired me to pursue this research area; hopefully continuing to bridge the gap between health care professionals and individuals with b-TM on the understanding of their lived experiences.

5.7 Concluding Thoughts

This research study used the qualitative methodology of interpretive phenomenology to gain a deeper understanding of the experiences of adolescent individuals with b-TM. Five research participants were recruited and interviewed to collect information on their lived experiences of living with, and managing b-TM on the cusp of transitioning from pediatric to adult institutions of care. This study identified that, although the experiences of adolescents with b-TM varied, reoccurring themes do exist. These include, 1) knowledge and understanding of b-TM; 2) familial support; 3) self-reflection and disclosure; 4) comradery; and 5) readiness to transition, including the impact of a *Health Transition Specialist*.

This research sheds light on the challenges encountered by adolescents with b-TM who anticipate transition and proposes important future areas for research. Transition to adult oriented institutions of care for adolescents with b-TM should be an on-going process, which can be facilitated by pediatric health care professionals to address the specific developmental needs of patients. Creating a collaborative transitional plan early on with the adolescent individual, their family, and health care team (including a *Health Transition Specialist*) which outlines a clear timeline for transition based on their development may help make the progression appear less subjective and help adolescents recognize the positive implications on their health, while also decreasing their apprehensions.

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Appendix A



Research Ethics Board (REB) Study Approval Letter

2017-07-19

Melanie Kirby-Allen
Haematology/Oncology

REB number: 1000056051

Study Title: Lost in Transition: Experiences of adolescent Beta-thalassemia Patients in Toronto

Date of Approval: 2017-07-19

Expiry Date: 2018-07-18

Thank you for the application submitted on **2017-05-26**. The above referenced study was reviewed through a delegated process (not by Full Board review). Any concerns arising from this review have been documented and resolved.

The REB voted to approve this study, and your participation as Principal Investigator, as it is found to comply with relevant research ethics guidelines, as well as the Ontario Personal Health Information Protection Act (PHIPA), 2004.

The Hospital for Sick Children Research Ethics Board hereby issues approval for the above named study. This approval is effective from **2017-07-19** to **2018-07-18**. Continuation beyond that date will require further review of REB approval.

The following document(s) have been reviewed and are approved:

- **Study Protocol, Version Dated: July 9, 2017 [REB1000056051 - STUDY AND RESEARCH PROPOSAL - CLEAN COPY - v07 09 2017.docx (1.0)]**
- **Interview Guide, Version Dated: March 2017 [Interview Guide - vMAR2017.docx (1.0)]**
- **Survey, Version Dated: June 21, 2017 [Thal Readiness Survey w NO IDENTIFIERS- v06 21 2017.pdf (1.0)]**
- **Consent Form, Version Dated: July 9, 2017 [Consent Form Letter of Invitation v07-09-2017 - CLEAN COPY.docx (1.0)]**

During the course of this investigation, any significant deviations from the approved protocol and/or unanticipated developments or significant adverse events should immediately be brought to the attention of the REB.

A handwritten signature in black ink, appearing to read "Kathy Boutis".

Kathy Boutis
REB Vice-Chair

555 University Avenue, Toronto, ON M5G 1X8
Tel: (416) 813-8279 Fax: (416) 813-6515



The SickKids REB operates in compliance with the Tri-Council Policy Statement; ICH Guideline for Good Clinical Practice E6(R1); Ontario Personal Health Information Protection Act (2004); Part C Division 5 of the Food and Drug Regulations; Part 4 of the Natural Health Products Regulations and the Medical Devices Regulations of Health Canada. The approval and the views of the REB have been documented in writing. The REB has reviewed and approved the clinical trial protocol and informed consent form for the trial. All investigational drug trials at SickKids are conducted by qualified investigators.

Furthermore, members of the Research Ethics Board who are named as Investigators in research studies do not participate in discussions related to, nor vote on such studies when they are presented to the REB.

Appendix B

Approval Notice - REB File #14289

Date: September 19, 2017
To: Caroline Barakat-Haddad
From: Jia Li, Acting REB Chair
File # & Title: 14289 - *Lost in Transition: Experiences of Adolescent Beta-thalassemia Patients in Toronto*
Status: **APPROVED** (Full REB approval granted as condition outlined in September 11, 2017 letter has been addressed)
Current Expiry: September 01, 2018

Notwithstanding this approval, you are required to obtain/submit, to UOIT's Research Ethics Board, any relevant approvals/permissions required, prior to commencement of this project.

The University of Ontario, Institute of Technology Research Ethics Board (REB) has reviewed and approved the research proposal cited above. This application has been reviewed to ensure compliance with the Tri-Council Policy Statement: Ethical Conduct for Research Involving Humans (TCPS2 (2014)) and the UOIT Research Ethics Policy and Procedures. You are required to adhere to the protocol as last reviewed and approved by the REB.

Continuing Review Requirements (all forms are accessible from the [IRIS research portal](#)):

- **Renewal Request Form:** All approved projects are subject to an annual renewal process. Projects must be renewed or closed by the expiry date indicated above ("Current Expiry"). Projects not renewed 30 days post expiry date will be automatically suspended by the REB; projects not renewed 60 days post expiry date will be automatically closed by the REB. Once your file has been formally closed, a new submission will be required to open a new file.
- **Change Request Form:** Any changes or modifications (e.g. adding a Co-PI or a change in methodology) must be approved by the REB through the completion of a change request form before implemented.
- **Adverse or Unexpected Events Form:** Events must be reported to the REB within 72 hours after the event occurred with an indication of how these events affect (in the view of the Principal Investigator) the safety of the participants and the continuation of the protocol (i.e. un-anticipated or un-mitigated physical, social or psychological harm to a participant).
- **Research Project Completion Form:** This form must be completed when the research study is concluded.

Always quote your REB file number (**14289**) on future correspondence. We wish you success with your study.

Dr. Jia Li
Acting REB Chair
jia.li@uoit.ca

Janice Moseley
Research Ethics Coordinator
researchethics@uoit.ca

NOTE: If you are a student researcher, your supervisor has been copied on this message.

Appendix C



RESEARCH ETHICS BOARD
OFFICE OF RESEARCH SERVICES

LETTER OF INVITATION

Title of Research Study: Lost in Transition: Experiences of Adolescent Beta-thalassemia Patients in Toronto

You are invited to participate in the research study named above. This study (#REB1000056051/#REB14289) has been reviewed by the Research Ethics Boards of The Hospital Sick Children and University of Ontario Institute of Technology. Before agreeing to participate in this study, it is important that you read and understand the benefits and risks of your participation in this research study. The following information describes the purpose, procedures, benefits, risks and precautions associated with this study. It also describes your right to refuse to participate or withdraw from the study at any time. To decide whether you wish to participate in this research study, you should understand enough about its risks and benefits to be able to make an informed decision. Please ask the study doctor or the study staff to explain anything you do not understand before signing this consent form.

Investigator(s):

Principal Investigator: Dr. Melanie Kirby-Allen-Kirby; Haematology/Oncology, Hospital For Sick Children, Associate Professor, University of Toronto.

Tel.: 416-813-7606 ext. 207606 **Email:** melanie.kirby-allen@sickkids.ca

Student Investigator: Julie Vizza MHSc. (Cand.) (Faculty Supervisor: Dr. Caroline Barakat-Haddad); Faculty of Health Sciences, University of Ontario Institute of Technology. **Tel.:** 905-721-8668 ext. 2173 **Email:** julie.vizza@uoit.net

Co-Investigator: Brooke Allemang; Health Transition Specialist; Sickle Cell & Thalassemia Programs; The Hospital for Sick Children. **Tel:** 416-813-7654 ext. 204722 **Email:** brooke.allemang@sickkids.ca

Purpose and Procedure:

You are being invited to participate in this study because you have beta-thalassemia major and you have indications that iron overload may be, or possibly will be, a problem for you in the future. Iron overload is caused by repeated blood transfusions. Sometimes, iron chelation treatment is needed because the human body cannot remove extra iron on its own. Also, sometimes, this excess iron may build up in the liver and/or the heart. This can cause health

problems. But if treated at an early stage, mild iron loading will cause no problems at all. The purpose of this study is to gain insight to your daily experiences living with and managing your thalassemia as an adolescent between the ages of sixteen and eighteen.

The research employs a short questionnaire and a semi-structured interview where you will be asked to discuss your experiences. The interviews are audio-recorded and will be transcribed verbatim; the researcher may also take notes by hand during the interview. It is anticipated that the questionnaire will take you 10-15 minutes to complete and the interview between 45 to 60 minutes to complete; therefore, participation in this study will take approximately 55 to 75 minutes. Following the completion of the recorded interview, it will be transcribed. A copy can be made available for you upon your request. This project requires a collaborative relationship of knowledge construction between you and the researcher. Therefore, should a subsequent meeting be necessary, to confirm meaning of statements and add necessary information, it will occur in person, on a different regularly scheduled clinic appointment date (approximately 30 minutes). This cyclical process may need to occur at your discretion to ensure that meaning has been accurately captured and your experience sufficiently described in as much detail as possible.

Potential Benefits:

You will not directly benefit from being in this study. However, information learned from this study may help other people with beta-thalassemia major in the future. That is, information obtained from this study may help your healthcare team better understand the experiences of individuals living with beta-thalassemia major, which may facilitate advancements in current healthcare practices and improve patient quality of life.

Potential Risk or Discomforts:

You may feel uncomfortable or embarrassed speaking about your experiences living with beta-thalassemia and your transition from a paediatric to adult institution of care. You do not have to answer any questions that you do not want to and can end the interview at any time should you feel uncomfortable for any reason. All data collected during the interviews are confidential and will only be accessed by the members of the research team listed on this consent form.

Storage of Data:

Recorded interviews will be transcribed manually from the audio recordings and may be shared with the research team members listed on this consent form. Audio recordings will be deleted, and any hand written notes taken by the principal investigator will be shredded, once transcribed. Please do not use the real names of yourself or anyone else during the interviews. Raw data transcripts will be destroyed after they have been analyzed. Additionally, in accordance to the Hospital for Sick Children policy, all data will be destroyed no later than 7 years from the date of individuals' participation in the study.

Confidentiality:

With your permission, your experiences as an adolescent beta-thalassemia patient will be collected for the purposes of informing the research questions and will only be shared with members of the research team listed on this form. The researchers will keep any personal health information about you in a secure and confidential location for the duration required by law. Additionally, a list linking your study number with your name will be filed separately in an alternate and secure location. A copy of your research consent form will be inserted in your health records. Confidentiality will be provided to the fullest extent possible by law, professional practice, and ethical codes of conduct. Additionally, representatives of The Hospital for Sick Children *Research Ethics Board* or *Research Quality and Risk Management* team may see your health record to check on the study. By signing this consent form, you agree to let these people review your records.

Right to Withdraw:

Your participation is entirely voluntary and whether you choose to participate or not will have no bearing on your current or future care at The Hospital for Sick Children. Additionally, you can choose to provide answers to only those questions that you are comfortable with. The information that is shared will be held in strict confidence and discussed only with the research team. You may withdraw at any time. If you withdraw, your data will be removed and destroyed and not included in the final research analysis. You do not need to disclose any reason for your request to withdraw your consent. There will be no negative consequences should you wish to withdraw from the study. Please note that it is not possible, to withdraw results once they have been published or otherwise disseminated.

Ethical Participant Concerns and Reporting:

If you have any questions concerning the research study, or experience any discomfort related to the study you are encouraged to contact any of the researcher(s) listed on this form. Any questions regarding your rights as a participant, complaints or adverse events may be addressed to Research Ethics Board at The Hospital for Sick Children at (416) 813-8279 or through the University of Ontario Institute of Technology Ethics Compliance Office (905) 721-8668 ext. 3693. By consenting, you do not waive any rights to legal recourse in the event of research-related harm.

Conflicts of Interest

The research team members of this study do not have any conflicts of interest to declare.

Debriefing and Dissemination of Results:

A hard copy of the published thesis research will be given to the Hospital for Sick Children library and information on how to access an electronic version of the thesis will also be communicated.

Patient Consent Form

Title: Lost in Transition: Experiences of Adolescent Beta-thalassemia Patients in Toronto

Consent to Participate:

By signing the consent form, I agree that:

- You have explained this study to me. You have answered all my questions.
- You have explained the possible harms and benefits (if any) of this study.
- I know what I could do instead of taking part in this study. I understand that I have the right not to take part in the study and the right to stop at any time. My decision about taking part in the study will not affect my health care at Sick Kids.
- I am free now, and in the future, to ask questions about the study.
- I have been told that my medical records will be kept private except as described to me.
- I understand that no information about who I am will be given to anyone or be published without first asking my permission.
- I freely consent to participate in the research study, understanding that I may withdraw my participation at any time without penalty.

I, _____, have read the information provided and any questions I have asked have been answered to my satisfaction. I agree to participate in this activity, realizing that I may withdraw at any time without reason and without prejudice (or where applicable - without prejudice to my future medical treatment).

I understand that all information provided is treated as strictly confidential and will not be released by the researchers. The only exception to this principle of confidentiality is if documents are required by law. I have been advised as to what data is being collected, what the purpose is, and what will be done with the data upon completion of the research. Also, I am aware that I will receive a copy of my signed consent form for my own records.

I agree that research data gathered for the study may be published provided my name or other identifying information is not used.

Participant Name (Print)	Participant Signature	Date (mm/dd/yyyy)
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The below signature acknowledges that I have explained the research study to the participant named above. I have answered all questions pertaining to the study to the satisfaction of the consenting participant.

Person obtaining consent (Print)	Signature	Date (mm/dd/yyyy)
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Appendix D

Copy of the “Thalassemia Readiness Survey”

SickKids®



HAEMATOLOGY/ONCOLOGY CLINIC 8D

Thalassemia Readiness Survey

EPC: Transition Readiness Assessment

Welcome to the readiness survey for the Thalassemia Program. The purpose of the survey is to help your health care team understand your readiness for transition to adult care. By completing this survey, your health care team can guide you in learning about the adult health care system. Your responses will be kept confidential and will only be shared with your health care team.

Please circle the most appropriate answer based upon your own health care knowledge.

1. I am:
 - a. Male
 - b. Female

2. I am:
 - a. 13 yrs old
 - b. 14 yrs old
 - c. 15 yrs old
 - d. 16 yrs old
 - e. 17 yrs old
 - f. 18 yrs old

3. I can describe my thalassemia to others.
 - a. Agree
 - b. Somewhat agree
 - c. Somewhat disagree
 - d. Disagree

4. I am in charge of preparing and taking my medication and/or treatments on my own.
 - a. Agree
 - b. Somewhat agree
 - c. Somewhat disagree
 - d. Disagree

5. Which medications do you take? Circle all that apply.
 - a. Folic acid
 - b. Deferasirox (EXJADE®)
 - c. Deferoxamine (DEFERAL®)
 - d. Calcium supplements
 - e. Vitamin D supplements
 - f. I don't know
 - g. None
 - h. Other _____

6. I know what my medications and/or treatments are for.
 - a. Agree
 - b. Somewhat agree
 - c. Somewhat disagree
 - d. Disagree
 - e. N/A

7. I know how my medications and/or treatments are paid for.
 - a. Agree
 - b. Somewhat agree
 - c. Somewhat disagree
 - d. Disagree

8. I can get myself to and from health care appointments on my own.
 - a. Agree
 - b. Somewhat agree
 - c. Somewhat disagree
 - d. Disagree
9. I know how to schedule a health care appointment on my own.
 - a. Agree
 - b. Somewhat agree
 - c. Somewhat disagree
 - d. Disagree
10. When I get sick, I know how to get the help I need.
 - a. Agree
 - b. Somewhat agree
 - c. Somewhat disagree
 - d. Disagree
11. I spend time alone with my health care provider at each visit.
 - a. Always
 - b. Sometimes
 - c. Rarely
 - d. Never
12. I talk to my health care provider about how thalassemia affects my life.
 - a. Agree
 - b. Somewhat agree
 - c. Somewhat disagree
 - d. Disagree
13. I know what my health condition may bring in the future.
 - a. Agree
 - b. Somewhat agree
 - c. Somewhat disagree
 - d. Disagree
14. Do you have a family doctor who you will continue seeing when you are an adult?
 - a. Yes
 - b. No
 - c. I don't know
15. What type of thalassemia do you have?
 - a. Alpha thalassemia major
 - b. Hemoglobin H
 - c. Beta thalassemia major
 - d. Thalassemia intermedia
 - e. Other
 - f. I don't know
16. What is your blood type?
 - a. A+
 - b. A-
 - c. B+
 - d. B-
 - e. O+
 - f. O-
 - g. AB+
 - h. AB-
 - i. I don't know
17. Do you know why you are scheduled to do the following tests: audiogram, ophthalmology, bone mineral density, echocardiogram, pulmonary hypertension screen, liver MRI, heart MRI?
 - a. Yes
 - b. No
 - c. I don't know
 - d. N/A
18. Are you up to date with your immunizations?
 - a. Yes
 - b. No
 - c. I don't know
19. I understand how using alcohol/tobacco/drugs affects my thalassemia and my medications.
 - a. Agree
 - b. Somewhat agree
 - c. Somewhat disagree
 - d. Disagree
20. I would feel comfortable asking my health care provider about sexual health and puberty.
 - a. Agree
 - b. Somewhat agree
 - c. Somewhat disagree
 - d. Disagree



HAEMATOLOGY/ONCOLOGY CLINIC 8D

Thalassemia Readiness Survey

LAST NAME	(FIRST)
MRN NUMBER	VISIT NUMBER
DATE OF BIRTH YYYY-MM-DD	SEX
ADDRESS	
IMPRINT OR ENTER DETAILS BY HAND	

21. I know where to find information about which methods of contraception (i.e. condoms, birth control pill, etc.) are right for me, given that I have thalassemia.
 a. Agree b. Somewhat agree c. Somewhat disagree d. Disagree
22. If I decide to start a family in the future, I understand how thalassemia may be passed on to my children since it is genetic.
 a. Agree b. Somewhat agree c. Somewhat disagree d. Disagree
23. How would you like information about thalassemia to be given to you? Circle all that apply.
- a. Pamphlet
 - b. Website
 - c. Video
 - d. Online tutorial
 - e. Presentation or workshop
 - f. Other _____
24. What is the best way for us to contact you to remind you of appointments? Circle all that apply.
- a. Phone call
 - b. Text message
 - c. Email

Please provide your phone number or email address below.

25. Is there anything you would like more information about regarding what happens after you leave SickKids?

26. What worries you most about leaving SickKids?

THIS SECTION IS FOR HEALTH CARE PROVIDERS TO COMPLETE

Given the results of this survey, the following topics were discussed with the patient today:

The following topics should be discussed with the patient in future clinic visits:

Print name

-MM-DD)