“DON’T LET IT HOLD YOU BACK” –
MEN, MASCULINITIES AND SEVERE HEMOPHILIA

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Abstract

Hemophilia is a congenital bleeding disorder that predominantly affects men. Prophylactic intravenous replacement of missing clotting factor, known as prophylaxis, is the most effective treatment method to prevent all internal bleeds caused by hemophilia. Despite availability of prophylaxis, it is not well understood why adult men with severe hemophilia still experience bleeding episodes resulting in irreversible joint damage and disability, particularly in adolescence and young adulthood. The purpose of this qualitative study was to explore men’s experiences of hemophilia across age groups and to explore the connections to masculinity in relation to men’s hemophilia management decisions. Adult men with severe and moderately severe hemophilia A (less than 2% factor levels) participated in three age stratified focus groups: 19 to 24 years old (n=3), 25 to 39 years old (n=4), and 40 years and older (n=4). Qualitative descriptive and constructivist grounded theory methods were used to analyze the data. Findings suggest that pain, and not earlier symptoms of a bleed, is the primary sign that prompts men to treat or seek medical help. Older men were most insightful about hemophilia and experienced the most physical and work limitations, particularly those who acquired viral infections through factor use. Many men reported experiencing a high number of injuries in adolescence, particularly in high school, a critical time period in the development of a man’s identity. Findings suggest that the process of learning to manage hemophilia extends into young adulthood and masculinities impact men’s experiences of severe hemophilia and hemophilia management decisions. Limitations imposed on men by hemophilia prompted them to rebel or protest against it by engaging in risky physical activities or delaying treatment regardless of perceptions of long-term health consequences. Men with hemophilia may benefit from education about early signs of bleeds and improved support in transitioning from adolescence
into adulthood. Some masculine ideals can be mobilized to optimize hemophilia management practices that could enable men to surmount limitations and achieve personal goals across their lifespan.
Preface

This thesis is unpublished, original work by the author, Laszlo Kalmar. I modified the qualitative study design and focus group guiding questions initially developed by Leanne M. Currie and Shannon Jackson to meet the needs of the study purpose. I completed the literature review, identified the research questions, collected the data (with John Oliffe), analyzed the data, and wrote the entire study with direction from my supervisor, Leanne M. Currie, and committee members John Oliffe, Shannon Jackson, and Deborah Gue. The research conducted in this study was approved by the University of British Columbia’s Research Ethics Board, certificate number: H12-02509.
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Dedication

I dedicate this thesis to the men with severe hemophilia who confronted the limitation of their disorder with courage, discipline and gratitude. A particular acknowledgement is due for men who acquired infectious diseases through factor products, for many this was a devastating life changing experience. I am honoured to have had the privilege to gain such an in-depth insight into the lives and experiences of the study participants.
Chapter 1: Background

1.1 Introduction

Hemophilia is an X-chromosome linked bleeding disorder in which a deficiency in factor VIII (Hemophilia A) or IX (Hemophilia B) results in internal bleeding episodes that are difficult to stop (Canadian Hemophilia Society, 2013; World Federation of Hemophilia, 2013). Females who have an affected X-chromosome are typically asymptomatic but are carriers of the disorder and have a 50 percent chance of passing the affected gene onto their children. Since males only have one X-chromosome, those who are affected are symptomatic and will pass the affected X-chromosome onto their daughters but not onto their sons (Hemophilia of Georgia, 2014). Therefore, hemophilia is considered to be a predominantly male disorder and females carry the gene (Canadian Hemophilia Society, 2013).

Bleeding into the major articulating joints (ankle, knee, elbow, shoulder, hip) are the most common internal bleeding episodes causing pain, swelling and loss of range of motion (Forbes, Aledort, & Madhok, 1997). Recurrent bleeds into joints can progress to irreversible joint damage resulting in disability (Canadian Hemophilia Society, 2013). Bleeds in muscles are also common but are less likely to have long term sequelae; and bleeds into the brain or other vital organs are rare but can result in death (World Federation of Hemophilia, 2013). Overall, recurrent internal bleeds result in lower quality of life, disability, and historically a shorter life expectancy (Aznar et al., 2012; Manco-Johnson et al., 2007; Tagariello et al., 2009; Zappa, McDaniel, Marandola, & Allen, 2012).

Hemophilia can be treated with the replacement of the missing clotting factor (called factor concentrate) (World Federation of Hemophilia, 2013). Infusing with factor concentrate intravenously after an active bleed, known as on-demand treatment, can relieve immediate
symptoms, but over time irreversible joint damage can still occur (Aznar et al., 2012). Administration of factor concentrate on a regular basis to prevent bleeding episodes, also known as prophylaxis, is the most effective method to decrease the risk of joint damage, minimize disability, and improve quality of life (Aznar et al., 2012; Zappa et al., 2012). Yet, just over half (55%) of Canadian adults with severe hemophilia A and 20% of those with severe hemophilia B receive prophylaxis and adherence rates decrease with age (Biss et al., 2008; Jackson et al., 2014; Zappa et al., 2012), (Dr. Shannon Jackson, personal communication, February 25, 2014). Furthermore, adolescent and young adult males have the lowest adherence rates to both prophylaxis and on-demand treatment and highest incidence of bleeding episodes (Zappa et al., 2012). It is not well understood why prophylaxis is not always adhered to among adults with hemophilia despite the long-term benefits (Lindvall, Colstrup, Loogna, Wollter, & Grönhaug, 2010; Llewellyn, Miners, Lee, Harrington, & Weinman, 2003; Remor, 2011).

Studies on men’s health issues have shown that socially constructed masculine ideals and behaviours influence men’s health and illness practices (Connell, 1995, 2005; Courtenay, 2000). For example, masculine ideals of strength, control, stoicism, and independence can prevent men from practicing health preserving behaviours. Masculine ideals are also constructed in multiple ways and some men may mobilize the aforementioned masculine ideals toward effectual self-management. Although masculinities have not been studied in the context of hemophilia, there are some indications that masculine ideals could play an important role in men’s hemophilia management decisions (Park, 2000). Therefore, in this study I explored the connections between masculinities and hemophilia.
1.2 Hemophilia

As described earlier, hemophilia is a rare, inherited bleeding disorder affecting mostly males which causes lower than normal clotting factor levels in the blood resulting in episodic internal bleeding (Canadian Hemophilia Society, 2013; World Federation of Hemophilia, 2013). These bleeds occur in relation to haemostatic challenges from surgery or invasive procedures, injury, or in some cases, no apparent cause. Based on the blood plasma level of the clotting factor, the disorder may manifest in mild, moderate or severe forms (World Federation of Hemophilia, 2013). Most people with severe and some with moderate hemophilia (Aznar et al., 2012) can experience bleeds due to minor trauma, strenuous or even minor repetitive physical activity (World Federation of Hemophilia, 2013). Bleeds most frequently occur in joints and surrounding muscles resulting in pain, bruising, swelling, and loss of range of motion. After several joint bleeds *chronic hemophilic arthropathy*, an arthritis-like disease, can develop in joints, most commonly in knees, ankles, and elbows resulting in physical disability (Aznar et al., 2012; Canadian Hemophilia Society, 2013; World Federation of Hemophilia, 2013; Zappa et al., 2012). For most older patients with severe hemophilia and chronic hemophilic arthropathy, the ability to engage in physical activities is significantly limited. Throughout their lives, men with severe hemophilia are cautioned to avoid strenuous or high impact physical activities (including vocations involving such activities) to reduce bleeding episodes, joint damage and disability (Canadian Hemophilia Society, 2013); however, some men attempt to pursue an active lifestyle. Men with severe hemophilia historically have had a lower quality of life and shorter life expectancy compared to the general population. (Canadian Hemophilia Society, 2013; Zappa et al., 2012).
1.2.1 Hemophilia Management

Replacing missing clotting factor with factor concentrate products is the standard treatment of hemophilia (Canadian Hemophilia Society, 2013; Lindvall et al., 2010; Manco-Johnson et al., 2007; World Federation of Hemophilia, 2013). Alternatively, mild hemophilia A can be treated with Desmopressin (a synthetic version of arginine vasopressin) which can help increase factor VIII levels (Franchini & Lippi, 2011). In addition, liver transplant could technically cure hemophilia as coagulation factors are produced in the liver; however, it is not pursued without other liver disease because transplant comes with its own lifelong challenges from requiring immunosuppressive therapy (World Federation of Hemophilia, 2013).

1.2.2 On-Demand Treatment

Administering the factor concentrate after a bleeding episode to rapidly stop the bleed is known as on-demand treatment (World Federation of Hemophilia, 2013). However, on-demand treatment on its own does not usually prevent the development of hemophilic arthropathy (Aznar et al., 2012; Manco-Johnson et al., 2007). Men with severe hemophilia using on-demand management strategies are likely to develop physical disability and have poorer quality of life than those on prophylactic treatment (Aznar et al., 2012).

1.2.3 Prophylaxis

The regular administration of clotting factor to prevent bleeding before it starts, by maintaining a therapeutic plasma activity of the clotting factor, is known as prophylactic treatment (World Federation of Hemophilia, 2013). Because the body removes about half of the clotting factor activity infused in every 12 to 24 hours, the frequency and dose of prophylaxis needs to be adjusted accordingly. For those who tend to bleed frequently, prophylaxis has been shown to be the most effective method to manage the disease because it substantially reduces
bleeding episodes (Aznar et al., 2012; Lindvall et al., 2012; Manco-Johnson et al., 2007). In addition, factor concentrate may be administered prophylactically prior to physical activity that is likely to cause a bleed (World Federation of Hemophilia, 2013). By reducing the number of bleeding episodes, prophylaxis significantly reduces joint damage, delays and even prevents development of disability, reduces hemophilia related hospitalization, and improves overall quality of life compared to those receiving on-demand treatment (Zappa et al., 2012).

1.2.4 Adherence to Prophylaxis

Several studies have explored patients’ adherence to prophylaxis (Fischer et al., 2001; Ho et al., 2014). According to a 2010 practice pattern survey in the United States, a majority of 3 to 12-year-old children received prophylaxis, but the use of prophylaxis dramatically decreased with age (Zappa et al., 2012). The largest number of patients switching from prophylaxis to on-demand treatment was among adolescents (13 to 17-year-old group) and young adults (18 to 24-year-old group) (Zappa et al., 2012). Additionally, adolescents and young adults experienced a high number of bleeding episodes and report low adherence rates to recommended treatment (Aznar et al., 2012; Zappa et al., 2012). Although the use of prophylaxis among adult patients has been increasing over the last decade, on-demand treatment remains the most common management method for men with severe hemophilia aged 25 or older, and on-demand treatment proportionally increases with age (Zappa et al., 2012). Considering the compelling evidence for prophylaxis treatment, the lack of adherence and high rates of bleeding episodes among adolescents and young adults is worrisome given that even minor bleeds or few bleeds can lead to irreversible joint damage (Manco-Johnson et al., 2007; Roosendaal, Jansen, Schutgens, & Lafeber, 2008). Therefore, despite having the disorder well-managed early on, adolescents and young adults risk irreversible joint damage and disability by switching to on-demand treatment,
even if only for short periods (Fischer et al., 2002; Manco-Johnson et al., 2007). Furthermore, despite evidence showing that on-demand is ineffective at preventing joint damage and disability (Aznar et al., 2012), adults with severe hemophilia still predominantly use on-demand instead (Zappa et al., 2012).

It is not well understood why prophylaxis is not utilized more among adults with severe hemophilia. Some reasons reported include: lack of knowledge about hemophilia, its symptoms, benefits of treatment, and long term consequences of not treating (Llewellyn et al., 2003; Remor, 2011; Zappa et al., 2012). However, men with severe hemophilia are the most knowledgeable about their disease and most likely to adhere to prophylactic treatment (Lindvall et al., 2010; Llewellyn et al., 2003; Remor, 2011). Therefore, these reasons seem to inadequately explain the low adherence to prophylaxis rates among adults with severe hemophilia. Other researchers have suggested practical reasons behind men’s decisions to use on-demand instead of prophylactic treatment including inconvenience, pain, interference with daily activities, and concern over vein access over time (Aznar et al., 2012; Lindvall et al., 2012; Lindvall et al., 2010; Remor, 2011). Furthermore, a potential complication of factor replacement is the development of antibodies, known as inhibitors, that destroy the factor before it could stop a bleed (World Federation of Hemophilia, 2013). Although inhibitors most commonly occur in children with severe hemophilia during the period of the first 75 exposures to factor products (World Federation of Hemophilia, 2013), some men avoid prophylactic treatment for fear of not being able to treat a major bleed if inhibitors develop (Lindvall et al., 2010).

Hemophilia can also affect men at a social and psychological level. Researchers report that some men are hesitant to talk about their disorder to others, including colleagues and employers. Some attempt to hide their bleeding episodes even from their loved ones. Some
refrain from keeping contact with others who have hemophilia (Lindvall et al., 2010). In addition, stigma surrounds hemophilia fueled by misconceptions about bleeding to death from a minor cut or blunt trauma (Remor, 2011). Lastly, socially constructed gender roles can influence men’s experience of hemophilia; for example, men with hemophilia experienced distress for not being able to play typical masculine sports, like rugby (Park, 2000). The gendered nature of these social and psychological impacts of hemophilia is poorly understood and needs to be explicitly studied.

1.3 Masculinity and Men’s Health

Given that hemophilia is a disorder that predominantly affects men, it is important to understand the disorder from the perspective of a masculinities framework (Connell, 1995). A masculinities framework suggests that diverse masculine behaviours influence men’s health and illness practices and therefore health outcomes. From the masculinities perspective, men who align to idealized masculinity are rewarded with dominant positions over others (women and weaker men), but dominance can be challenged at any moment (Connell, 1995). Therefore, men are pressured to constantly construct masculine ideals to gain and maintain dominance. However, researchers have found that men’s alignments to masculine ideals can influence their overall health and life expectancy (Connell, 2005; Courtenay, 2000). Historically, masculine practices have been touted as hindering men’s ability to look after their health, and pursuing dominance and power may expose them to even greater health risks (Connell, 2005; Courtenay, 2000; Stanistreet, 2005). For example researchers found that men with heart disease who align to Western masculine ideals of strength and independence were particularly hesitant to seek help for chest pain because of fear of being seen as weak and failing to fulfill dominant social constructions of masculinity (Galdas, Cheater, & Marshall, 2007; Robertson, Sheikh, & Moore,
Yet, this behaviour placed men at high risk for heart attacks and heart disease. Similarly, men with depression tend to hide their feelings, avoid talking about their emotions and try to handle it on their own to avoid being perceived as weak and vulnerable (Emslie, Ridge, Ziebland, & Hunt, 2006; Oliffe & Phillips, 2008). By not talking about their feelings men can become irritable, aggressive, and be at risk for suicide (Oliffe & Phillips, 2008).

However, some men proactively engage in health preserving activities or seek medical help when needed, particularly if it allows them to embody manly virtues, such as strength, sexual performance, and being a good provider and protector (O'Brien, Hunt, & Hart, 2005). Bodily changes caused by prostate cancer and its treatments (e.g., erectile dysfunction, incontinence, weight gain) and not being able to work and needing to rely on others for help can threaten men’s masculine identities (Cecil, McCaughan, & Parahoo, 2010; Dieperink, Wagner, Hansen, & Hansen, 2013; Oliffe, 2005). However, men with prostate cancer can be motivated to seek medical help in order to preserve their sexual performance, ability to provide for their family, and maintain their social capital (e.g., power, success) (Dieperink et al., 2013). Furthermore, in contrast to Caucasian men, UK immigrant men of Asian origin were found to seek medical help much sooner for heart disease related symptoms (Galdas et al., 2007). These findings support the concept that men evaluate their health needs in the context of their male identity and perceived social expectations, both of which connect to culture and socioeconomic status.

Masculinities in the context of hemophilia has not been studied. However, Park (2000) found that men with hemophilia in New Zealand often described being unable to play rugby as a big loss in their lives. This is significant if we understand that in New Zealand playing rugby is
consistent with masculine ideals of strength, courage, honour and discipline. Playing rugby is an important part of developing and constructing a male identity; participating in other non-contact sports can be a poor substitute in meeting those ideals. For many, the immediate reward of playing rugby (fitting in, being a man) was much stronger than the fear of physical disability (Park, 2000). The findings by Park (2000) suggest that men need to reconcile their hemophilia management choices not just with their knowledge of the disorder and risk for injuries, but also with their immediate social and psychological needs and pressures.

1.4 Problem Statement

Despite evidence showing that prophylaxis is the best method of managing severe hemophilia by preventing bleeds in the first place (Aznar et al., 2012), about half of adult men with severe hemophilia A use prophylaxis (Biss et al., 2008; Zappa et al., 2012). Some studies suggest that lack of knowledge may contribute to why prophylaxis is not utilized more among men with severe hemophilia (Llewellyn et al., 2003; Remor, 2011). Other studies suggest that men with severe hemophilia are the most knowledgeable (Lindvall et al., 2010), and those who bleed more frequently are wiser about the consequences of not treating (Llewellyn et al., 2003; Remor, 2011). This discordant relationship however does not explain the lack of adherence. However, Lane et al. (2013) suggests that timing, rapport, and context can determine individuals’ readiness to uptake and assimilate information into hemophilia management practices. Additionally, other research indicates that masculine roles play an important part in men’s hemophilia management decisions (Park, 2000). In other men’s health issues, research indicates that socially constructed masculine ideals impact health related behaviours (Courtenay, 2000). Therefore, in order to better understand the factors behind poor utilization of prophylaxis among
men with hemophilia, it is important to further explore their experiences of hemophilia and the connections to masculinity in relation to men’s self-management decisions.

1.5 Purpose

The purpose of the study was to explore men’s experiences of severe hemophilia through a wide lens and describe the connections between socially constructed masculinities and hemophilia management decisions. The findings can assist with better understanding the contexts in which men manage their disorder. A comprehensive understanding of men’s experiences can be used to guide improvements in hemophilia care services and to further advance research in this field.

1.6 Significance

Despite advances in hemophilia care, life expectancy and quality of life of men with severe hemophilia remains lower than the general population (Aznar et al., 2012). Research shows that with prophylaxis, bleeding episodes can be reduced resulting in fewer disabilities, number of hemophilia related surgeries amid improved quality of life and increased life expectancy (Aznar et al., 2012). However, many adolescent and young adult patients switch from prophylaxis to on-demand treatment resulting in increased bleeding episodes (Zappa et al., 2012). Additionally, the use of prophylaxis decreases with age and only about half of all adults with severe hemophilia A use prophylaxis (Biss et al., 2008; Zappa et al., 2012). In order to further improve hemophilia care and the health of men, we need to better understand the connections between men’s hemophilia and masculinities.

1.7 Research Questions

Research Question #1

How is severe hemophilia experienced among men of varying age groups?
Research Question #2

What are the connections between masculinities and men’s experiences of severe hemophilia across varying age groups?
Chapter 2: Literature Review

2.1 Introduction

In this chapter hemophilia is described including its prevalence, common symptoms, management strategies, and adherence to treatment among adult males. Literature was also explored for barriers to adherence to recommended treatment among adult males. In addition, the literature on masculinity in the context of health is summarized to reflect the current state of the science and how masculinities are interrelated with men’s health practices, particularly in the context of heart disease, prostate cancer, and depression. Finally, the connections between masculinities and hemophilia described in the literature are reviewed.

2.2 Literature Search

CINAHL, PubMed, and EMBASE databases were searched for the most recent literature on hemophilia. The search terms used included “hemophilia”, “hemophilia AND management”, “hemophilia AND treatment”, and “hemophilia AND prophylaxis”. Only scholarly articles with male subjects published between 2003 and 2013 were considered totaling over 300 articles which were then further screened for relevance of hemophilia management, treatment, prophylaxis, exercise, and men’s adherence to treatment. Reference lists of publications were reviewed for additional primary key articles on hemophilia. Finally, online resources and books were utilized to gain information on other relevant aspects of hemophilia and its management.

On the topic of masculinities, several search strategies were also used to conduct a wide-ranging literature review. First, a preliminary search was completed using terms: “men, masculinities and health” in Google Scholar to gain an understanding of the types of literature available. In CINAHL, the search terms: “men masculinity AND health” were used for scholarly journals published between 1994 and 2013 which resulted in 195 articles. Further limiting the
literature to male subjects only and abstracts available resulted in 162 articles which were further screened for relevance to men’s masculinity(ies) and health including heart health, cancer (prostate, penile, colon), depression, and smoking cessation as well as having been conducted in Canada, North America, Western Europe and Australia. This screening process resulted in 24 articles. Modifying the search terms to “men and masculinities and health” and completing the same screening process resulted in an additional 8 articles, for a total of 32 articles. Using the same search terms in PubMed and EMBASE databases resulted in the same articles as found in CINAHL.

None of the articles found up to this point addressed masculinity(ies) and hemophilia. Therefore, I proceeded to use search terms “men AND masculinity AND hemophilia”, “men AND masculinities AND hemophilia”, “hemophilia AND masculinity” and “hemophilia AND masculinities” but all returned 0 results. The search term “men AND hemophilia” returned 17 results when limited to scholarly articles published between 1993 and 2013 and to male subjects only.

Both in PubMed and in EMBASE, I retrieved a few additional articles on masculinities. However, using “related articles” in CINAHL, PubMed and/or EMBASE and checking the reference lists of selected articles for key references resulted in finding additional primary literature. Overall, the literature on masculinities provides well developed knowledge based on qualitative studies. Additionally, there were well established literature and discourse on theory of masculinities (Connell, 2005; Courtenay, 2000). However, there was no literature that addressed masculinities in the context of hemophilia. This was an important knowledge gap in the literature which I argue later in this chapter will be addressed in part by this study.
2.3 Hemophilia

2.3.1 Prevalence and Heredity

Hemophilia is a recessive congenital coagulation disorder where a mutation in the clotting factor genes causes deficiency in clotting factor VIII (known as Hemophilia A) or factor IX (known as Hemophilia B) (World Federation of Hemophilia, 2013). Hemophilia is a rare disorder; hemophilia A occurs in about 1 in 10,000 births (World Federation of Hemophilia, 2013) and 2,966 Canadians have hemophilia A (Association of Hemophilia Clinic Directors of Canada, 2013). Hemophilia B is even more rare, affecting about 1 in 50,000 births worldwide and 691 Canadians (Association of Hemophilia Clinic Directors of Canada, 2013).

The genetic mutation(s) that cause hemophilia A and B are on the X-chromosome (World Federation of Hemophilia, 2013). Females who carry hemophilia typically have one affected X-chromosome and have a 50% chance of passing it on to their children (World Federation of Hemophilia, 2013). Some females can experience a milder form of the disorder because the healthy X-chromosome can still initiate the production of the factor, even if not to the desired level (World Federation of Hemophilia, 2013). Severe hemophilia in women is extremely rare, but can occur if both X-chromosomes are faulty or the healthy X-chromosome is inactivated leaving the faulty X-chromosome controlling factor production (Canadian Hemophilia Society, 2013). On the other hand, males are typically symptomatic if their only X-chromosome is affected and will pass the affected X-chromosome onto their daughters but not onto their sons (Canadian Hemophilia Society, 2013). Therefore, hemophilia is a predominantly male disorder and females are carriers of it (Canadian Hemophilia Society, 2013).

Hemophilia is also known as “The Royal Disease” because it affected several European royal families, including British, Spanish, German, and Russian. It is suggested that Queen
Victoria of England was a carrier and passed on the disorder to other Royal families through the marriages of her off-spring (Canadian Hemophilia Society, 2013). However, hemophilia can be found in all ethnic groups (Canadian Hemophilia Society, 2013). Although, most hemophilia is passed on through an affected X-chromosome from the mother or father, up to a third of all cases occur due to spontaneous mutation of the coagulation gene at the time of conception in a male or female fetus (Canadian Hemophilia Society, 2013; World Federation of Hemophilia, 2013).

2.3.2 Clinical Symptoms

The deficiency in clotting factors prevents the completion of the coagulation cascade and therefore the blood clot forms more slowly than normal (Canadian Hemophilia Society, 2013). Depending on the serum level of the affected clotting factor, hemophilia is categorized as: severe (<1% of normal), moderate (1-5% of normal) and mild (5-40% of normal) (World Federation of Hemophilia, 2013). Men with mild hemophilia usually bleed only as a result of a trauma or surgery (World Federation of Hemophilia, 2013). With moderate hemophilia, bleeds are caused by surgery, minor trauma, strenuous physical activity, or playing contact sports but can also appear to be spontaneous. Some men with moderate hemophilia rarely bleed, but a small number of them have a similar bleeding tendency as men with severe hemophilia. Severe hemophilia has the most extreme impact on a man’s everyday life because frequent and apparent spontaneous bleeding episodes cause pain, loss of range of motion, disability and limit physical activity (Canadian Hemophilia Society, 2013; World Federation of Hemophilia, 2013). However, 10-15% of men with severe hemophilia have a milder bleeding phenotype than predicted based on factor level due to genetic factors outside of the FVIII or FIX gene (Aznar et al., 2012). In addition to trauma and surgery, men with severe hemophilia may bleed due to physical activity such as lifting (causing muscle tears and bleeding), repetitive impact on joints.
(e.g., running or skating), or contact sports (e.g., football and hockey where men are exposed to
minor traumas, strain on joints, or twisting of joints) (World Federation of Hemophilia, 2013).
Bleeds that have no obvious trigger are often described as ‘spontaneous bleeds’ and are
characteristic of severe hemophilia (Canadian Hemophilia Society, 2013; World Federation of
Hemophilia, 2013); however, spontaneous bleeds are more likely to be caused by minor
repetitive physical activities, including typing or writing (Roosendaal et al., 2008).

Bleeding into a joint is the most common form of internal bleeding, including bleeding
into ankle, knee, elbow, shoulder, and hip (Aznar et al., 2012; Hemophilia of Georgia, 2014).
Bleeds into a joint can lead to joint inflammation (hemophilic synovitis), cartilage damage, bone
loss, mobility problems, loss of range of motion, and development of hemophilic arthropathy (an
arthritis like joint disease) (Canadian Hemophilia Society, 2013; Manco-Johnson et al., 2007;
Zappa et al., 2012). Bleeds into muscles are less common and are typically resolved without
long term effects. However, bleeds into certain muscles left untreated can destroy the muscle,
damage nerves or cut off circulation to the lower part of an extremity resulting in loss of function
(Hemophilia of Georgia, 2014). Bleeding into vital organs, including the brain, can result in
death (Canadian Hemophilia Society, 2013). Men with hemophilia are also at increased risk for
renal disease due to renal bleeding episodes, viral infections, drug nephrotoxicity, and possibly
high protein load from factor replacement (Esposito et al., 2013). Adults with severe hemophilia
can be physically disabled due to the progression of hemophilic arthropathy in their middle or
older age (Zappa et al., 2012).

For the most part, hemophilia A and hemophilia B are considered to have similar clinical
presentations. However, research suggests that hemophilia B tends to have a milder bleeding
phenotype resulting in a different disease process and presentation compared to hemophilia A
Despite similar replacement factor use (approx. 105,000 IU/patient/year) among patients with comparable severity of the disease, there are more frequent bleeding episodes among patients with hemophilia A than with hemophilia B (Nagel, Walker, Decker, Chan, & Pai, 2011). Furthermore, patients with hemophilia A proportionately undergo surgical procedures for musculoskeletal complications three times more often than patients with hemophilia B (Nagel et al., 2011; Tagariello et al., 2009). These findings suggest, on average, a more severe disease process among patients with hemophilia A.

2.4 Hemophilia Management

In Canada, prior to the 1950s, treatment of hemophilia mostly consisted of rest, pressure dressings, ice, and cauterization. Individuals with severe hemophilia didn’t live to adulthood due to intracranial bleeds or severe bleeds after a major injury or surgery (Canadian Hemophilia Society, 2013). Males with severe or moderate forms of hemophilia who grew into adulthood were crippled with severe joint disease and had a significantly shorter lifespan (Canadian Hemophilia Society, 2013). Routine replacement of the missing clotting factor started in the 1950s with transfusing first with whole blood then with fresh-frozen plasma, but limiting the treatment to only hospital settings (likely treating only life threatening bleeds) delayed treatment and reduced its effectiveness. It was the development of cryoprecipitate, a plasma fraction, in 1964, that resulted in significant improvements in quality of life and life expectancy (Pemberton, 2011). Commercially produced factor concentrates soon followed and these factor concentrates made home infusions feasible in the 1970s (Pemberton, 2011). Home infusion is the intravenous administration of the factor concentrate by a parent, other care giver, or the person with hemophilia in the individual’s every-day setting (i.e., home, work, etc.). This was a dramatic advancement in hemophilia care as it made treatment available immediately after the start of a
bleed and resulted in enhanced quality of life, decreased mortality rates, and increased life expectancy compared to previous generations of males with hemophilia (Pemberton, 2011).

Unfortunately, the same advancements in hemophilia care resulted in dire consequences in the 1980s; approximately half of all men with hemophilia and 90 to 95 percent of men with severe hemophilia acquired blood-borne infectious diseases, including HIV and Hepatitis C, through factor replacement therapy resulting in debilitating diseases and premature death (Pemberton, 2011). These consequences occurred because of the human plasma origin of the products and the lack of safety requirements and testing for blood-borne viruses (Pemberton, 2011). The chance of contracting these viral infections was significantly higher for men who used prophylaxis compared to those who used on-demand treatment because of the increased exposure to factor concentrate. For this reason, prophylactic treatment using plasma derived factor concentrate for the most part was abandoned (Pemberton, 2011).

Recombinant factor, a genetically engineered factor concentrate was developed in the 1990s by cloning the clotting gene (Pemberton, 2011). In Canada, recombinant factor VIII (FVIII) became available in 1993 and recombinant factor IX (FIX) in 1998 (Canadian Hemophilia Society, 2013). Because recombinant factor concentrate was not derived from human plasma, the risk of spreading infections was all but eliminated (Manco-Johnson et al., 2007). The development of a safer factor replacement concentrate made prophylaxis again a feasible method to manage hemophilia. However, because of the earlier devastating negative consequences of prophylactic use of factor concentrates, the uptake of prophylaxis as primary treatment method was slow among men with hemophilia (Pemberton, 2011).

In order to minimize the number of bleeding episodes, clinicians have historically suggested limiting physical activity, especially for those with severe hemophilia (Czepa, von
However, recent research indicates that inactivity results in weaker muscles around the joints potentially exposing the joints to higher risk of injuries (Czepa et al., 2013). Regularly engaging in low impact and controlled physical activity is now considered to be beneficial for patients with hemophilia (Canadian Hemophilia Society, 2013). Regular low impact exercise results in individuals’ self reporting of feeling better and only minor observable improvements on joints and physical conditions. However, some researchers recommend including exercise as an integral part of hemophilia treatment programs (Czepa et al., 2013). However, more studies are required to better understand the impact of increased activity levels on long-term patient outcomes (Czepa et al., 2013).

Today, there is still no cure for hemophilia (Pemberton, 2011). Despite significant advancement in hemophilia care over the last few decades, men with hemophilia may have to face lower socioeconomic status, lower quality of life, higher mortality rate, and shorter life expectancy when compared to the rest of the population because of the chronic health complications of hemophilia (Zappa et al., 2012). The standard of care for hemophilia includes intravenous replacement of the missing clotting factors (Canadian Hemophilia Society, 2013; Lindvall et al., 2010; Manco-Johnson et al., 2007; World Federation of Hemophilia, 2013). However, the time and frequency of the treatment varies depending on severity of the disorder, frequency of bleeds, (World Federation of Hemophilia, 2013), cost of treatment (Lindvall et al., 2012), and possibly because of the preference of the health care provider (Lindvall et al., 2010).

2.4.1 On-Demand Treatment

Administering the replacement clotting factor after a bleeding incident to stop the bleed is called on-demand treatment (World Federation of Hemophilia, 2013). In other words, factor replacement is only administered when symptoms indicate an actual bleed. The goal of
administering the replacement clotting factor immediately after the bleed is to stop the bleed as quickly as possible, reduce joint pain, minimize swelling, and restore joint mobility that results in improved quality of life for the patient (Aznar et al., 2012). On-demand treatment is recommended for patients with milder bleeding phenotype, typically patients with mild or moderate hemophilia (Nagel et al., 2011; Zappa et al., 2012). There are no standardized guidelines on how to administer on-demand treatment (e.g., timing, dosage) (Zappa et al., 2012). However, some research shows that on-demand treatment is not effective in preventing the development of hemophilic arthropathy in patients with severe hemophilia of high bleeding tendency (Aznar et al., 2012).

2.4.2 Prophylaxis

The regular infusion of clotting factor to prevent bleeds in the first place is known as prophylaxis (World Federation of Hemophilia, 2013). For individuals with moderate to severe forms of the disorder with high bleeding tendency, administration of the factor on a regular basis is found to be the most effective treatment method (Lindvall et al., 2012; Manco-Johnson et al., 2007; Zappa et al., 2012). Primary prophylaxis is the regular continuous treatment started before the second large joint bleed and age of 3 years (World Federation of Hemophilia, 2013). Secondary prophylaxis is the regular continuous treatment started after 2 or more large joint bleeds but before the onset of joint disease (World Federation of Hemophilia, 2013). Primary and secondary prophylaxis is applied to a greater extent in pediatric patient populations to protect the joints at an early stage (Zappa et al., 2012). Tertiary prophylaxis is the regular continuous treatment started after the onset of joint disease (hemophilic arthropathy) to prevent further damage (World Federation of Hemophilia, 2013) and it is typically used to prevent recurrent bleeds in joints that are prone to frequent re-bleeding (Zappa et al., 2012). Intermittent
prophylaxis is the preventive administration of replacement factor for a short period of time prior to an activity or event that is likely to cause a bleed (e.g., sporting event, surgery) (World Federation of Hemophilia, 2013). Frequency and dose of prophylactic treatment is not standardized, rather established based on individual needs and may range from daily, two or three times a week, or weekly administration (Canadian Hemophilia Society, 2013; Lindvall et al., 2012). The literature is inconsistent in the definition of prophylaxis but for the purpose of this study the definition of prophylaxis by the World Federation of Hemophilia was used as outlined above.

Research indicates that patients who receive prophylactic treatment, particularly primary and secondary prophylaxis, have fewer hospitalizations, fewer hemophilia related surgeries, and overall better quality of life compared to those receiving on-demand treatment only (Zappa et al., 2012). Prophylaxis is predominantly used in the 3 to 12-year-old patient population, but on-demand treatment remains the primary treatment method in the adult population (Zappa et al., 2012). Applying tertiary prophylaxis treatment in adult patients with irreversible joint damage can slow the progression of the disease and improve overall quality of life (Aznar et al., 2012). Accordingly, the use of prophylactic treatment has been increasing and the use of on-demand treatment has decreased in recent years among men with severe hemophilia (Zappa et al., 2012).

It is important to emphasize that subclinical bleeds (undetected chronic micro-haemorrhages within joints) and even a few bleeds can lead to long term joint damage (Fischer et al., 2002; Manco-Johnson et al., 2007; Roosendaal et al., 2008; Zappa et al., 2012). Using MRI examinations, Manco-Johnson et al. (2007) found irreversible joint damage that was not detected on physical examination or radiography studies in boys with severe hemophilia. Some boys with joint damage on MRI examination had no bleeding episodes during the study, and over half of
them experienced fewer than 10 bleeds. Additionally, prophylaxis reduced joint damage by 83% compared to on-demand treatment. Therefore, prophylactic treatment is an important strategy in preventing bleeds and long term joint damage (Manco-Johnson et al., 2007).

Prophylaxis results in better quality of life compared to on-demand treatment for men with severe hemophilia, but it also costs substantially more than on-demand treatment due to the increased consumption of factor replacement products (Carlsson et al., 2004; Manco-Johnson et al., 2007). Additionally, the health care costs for hemophilia are estimated to be 20 to 30 times of the general population with chronic illnesses (Johnson & Zhou, 2011). To gain some sense of the figures, prophylactic treatment for a child weighing 50 kg is estimated at $300,000 per year (in 2006 US dollars) (Manco-Johnson et al., 2007). The high cost of factor replacement therapy can make optimal care out of reach for some men in countries that don’t have a universal health care system (i.e., the United States) and even more so in developing countries (Johnson & Zhou, 2011).

Men with hemophilia in Canada are not directly exposed to the high cost of treatment because the universal health care system funds treatment. It is the provincial governments that ultimately pay for treatment through the health care agencies and are directly impacted by the cost. Policy makers on behalf of the provincial governments need to take into account not just the high cost of factor replacement but also the direct and indirect cost of illness (i.e., orthopedic surgeries on damaged joints, loss of productivity, early mortality) and the short and long-term quality of life, (Johnson & Zhou, 2011). Standard prophylactic treatment for hemophilia A ranges from two infusions per week to daily infusions. Although daily prophylaxis may appear to be the costliest treatment regime due to the frequency of infusions, in some cases it can result in about 30% reduction of the overall cost of hemophilia treatment by preventing bleeds,
avoiding on-demand treatment and joint surgeries (Lindvall et al., 2012). However, some men with hemophilia experience daily prophylaxis as more burdensome if venous access is difficult or the time involved interferes with daily life (Lindvall et al., 2012).

2.4.3 Inhibitors

The immune system of a person with hemophilia may develop antibodies, also known as inhibitors, against the FVIII or FIX protein which neutralize the factor before it can stop a bleed (World Federation of Hemophilia, 2013). Children with severe hemophilia (under 9 years of age) most commonly develop inhibitors and these occur during the period of the first 75 exposures to factor products (World Federation of Hemophilia, 2013); however, inhibitor development may occur at any age (Webert et al., 2012). The prevalence of inhibitors among Canadian patients with severe hemophilia A and severe hemophilia B is 8.9% and 2.6% respectively (Webert et al., 2012).

The development of inhibitors results in either ineffective treatment, or in some cases, a requirement for high doses of the factor concentrate to overwhelm the inhibitor for the same therapeutic effect. However, immune tolerance induction, the regular and sustained administration of the factor concentrate in large doses, can be utilized to reduce the antibody production after several months of treatment (World Federation of Hemophilia, 2013). With application of the immune tolerance induction technique, inhibitors can typically be resolved in about a year for approximately 80% of cases (Webert et al., 2012). Alternate clotting factors (without significant amounts of the antigen), also known as bypassing agents, can be used to treat a major bleed, but they are costlier and not as widely available (Webert et al., 2012). Alternatively, in some jurisdictions, plasmapheresis, a procedure that results in the rapid reduction of protein inhibitors present in plasma, can also be considered prior to administering
the factor concentrate (World Federation of Hemophilia, 2013). Treatment of inhibitors can be difficult, costly, may take a long time, and requires specialized medical expertise that only hemophilia treatment centers can offer. In addition, inhibitor development significantly increases the disease burden for the individual because of increased frequency of treatments or difficulty of stopping bleeds (World Federation of Hemophilia, 2013).

2.4.4 Adherence to Treatment

Despite evidence clearly indicating the benefits of treating with factor concentrate, researchers report varying adherence rates to both on-demand and prophylactic treatment among men with hemophilia (Remor, 2011). Although it is difficult to gain a clear picture of the exact adherence rate due to limited means to obtain accurate information, literature indicates that prophylaxis is used at the highest rate among patients 3 to 12 years of age (Zappa et al., 2012). However, switching from prophylaxis to on-demand is highest among patients 13 to 24 years of age despite high bleeding incidence and low adherence to treatment rates among the same age group compared to other age groups (Zappa et al., 2012). Adherence with prophylactic treatment declines in adolescence and young adulthood (Aznar et al., 2012).

Lack of knowledge about hemophilia, benefits of treating with factor concentrate, and adverse consequences of not treating have been frequently reported as reasons for not adhering to recommended treatments (Aznar et al., 2012; Llewellyn et al., 2003; Remor, 2011; Zappa et al., 2012). Furthermore, research shows that the more hemophilia-related symptoms men experience and the more knowledgeable they are of the need to treat, they are more likely to adhere to the prophylactic treatment regime in order to avoid exacerbation of symptoms (Llewellyn et al., 2003). Knowledge about hemophilia is the lowest among men with mild hemophilia (Lindvall et al., 2010) and men with severe hemophilia have the least difficulty with treatment (Lindvall et
it is suggested that more knowledge of adverse consequences and benefits of treatment are required for all men with hemophilia regardless of severity of the disease (Lindvall et al., 2012; Lindvall et al., 2010).

Other, more practical aspects of everyday life can also act as barriers to adhering to recommended treatment routines, especially when applying prophylactic infusion without actual bleeding episodes. Infusing factor concentrate on a daily basis can be time consuming, inconvenient, and interfere with daily activities (Aznar et al., 2012; Lindvall et al., 2012; Remor, 2011; Zappa et al., 2012). In addition, daily prophylactic treatment increases the frequency of venipunctures, raises concern regarding long-term vein access, and can cause pain, discomfort, and lower quality of life (Lindvall et al., 2012). Others are hesitant to follow prophylactic treatment routines for fear of developing inhibitors or virus transmission from the treatment (Lindvall et al., 2010; Remor, 2011).

As stated earlier, a large number of adolescents and young adults who previously received prophylactic treatment switch to on-demand treatment and experience high number of bleeding incidences (Zappa et al., 2012), but the reasons for this phenomenon are not well understood. However, this is a particularly troubling finding considering that even subclinical bleeds or fewer than 10 bleeds can result in joint damage (Manco-Johnson et al., 2007). In addition, researchers report that while most men with hemophilia inform their friends about their disorder (94%), fewer share that with their colleagues and employers (76%) (Lindvall et al., 2010). Yet, informing the workplace of the disorder could improve workplace safety. In addition, less than half of patients diagnosed with hemophilia keep contact with others with the same disorder (Lindvall et al., 2010). Also, both adults and children may attempt to hide injuries from their family until the symptoms become severe (Lindvall et al., 2010). Furthermore,
stigmatization can surround hemophilia in settings where others are not familiar with the disorder (Remor, 2011). Lastly, some men reported that a daily prophylaxis regime reminded them too much of their disorder (Lindvall et al., 2012). Together these findings reveal important social and psychological aspects of men’s experiences of hemophilia and their management decisions that are not well understood.

2.5 Masculinity

Statistics show that in Canada as well as in the majority of the developed world, life expectancy for men is lower than for females (Gough, 2013; Phillips, 2006; Sloan, Gough, & Conner, 2010; Statistics Canada, 2012). This phenomena has intrigued health researchers for decades and fueled much of early men’s health research. Although there are multiple perspectives about why the gender difference in life expectancy exists, Connell’s *Masculinities* (1995, 2005) framework has prevailed to evaluate connections between gender and men’s health outcomes. Connell identifies four stratifications within the masculinities: i) hegemonic masculinity, ii) complicit masculinity, iii) subordinate masculinity, and iv) marginalized masculinity. *Hegemonic masculinity* is a socially constructed ideal form of masculinity whereby men have dominance or power over women and other men. *Hegemonic masculinity* is represented only by a small number of men that produce and reproduce specific socially constructed behaviours and ideals. In other words, *hegemonic masculinity* represents the socially determined behaviour that claims male dominance. Western societies emphasize autonomy, control, stoicism, self-reliance, and toughness as hegemonic masculine attributes. The term *complicit masculinity* identifies that although only a very few men are in dominant positions within society, many men are complicit in sustaining hegemonic masculinity, and they benefit from the inequalities within society by being dominant over women and subordinating or
marginalizing other men. Subordinate masculinity is a less powerful form of masculinity than hegemonic or complicit masculinity visible through lower socio-economic conditions, and often stigmatized or excluded from the mainstream society. Lastly, marginalized masculinity refers to men whose full participation is limited due to material and structural constraints, such as race (i.e., being a visible minority) (Connell, 2005). It is also suggested that perceptions of masculinities are dependent on social contexts as some behaviours may be perceived as dominant (hegemonic) in some situations but subordinate or marginalized in other situations (Creighton & Oliffe, 2010).

In Connell’s framework he posits that diverse masculine behaviour influences men’s health and illness practices and therefore health outcomes. However, masculine practices often run counter to health preserving practices (Courtenay, 2000). Connell (2005) identified protest masculinity that occurs when men without masculine capital overemphasize toughness and risk-taking and engage in self-destructive behaviour in order to claim purchase on hegemonic masculinity. For example, marginalized men may risk opiate overdose (Stanistreet, 2005) or gay men may disregard the risks of having unprotected sex with a high number of sexual partners (Connell, 1995) in order to prove their masculinity and protest their position within masculine hierarchies.

Courtenay (2000) argued that socially required behaviours to construct and enact masculinity or femininity impacts one’s overall health. Health related beliefs, behaviours, and practices can be utilized to embody masculinities through language, work, sports, crime, and sex. Demonstrating dominant masculine ideals of independence, self-reliance, strength, and toughness can garner power for men. However, such behaviours can put men at a higher risk for health related problems and a shorter life expectancy (Courtenay, 2000). In other words, some
men engage in self-harming practices in the pursuit of masculine power. These behaviours can manifest as denying pain, weakness and vulnerability, maintaining emotional and physical control, displaying aggressive behaviours and exerting dominance (Courtenay, 2000). Tolerating injuries is a common masculine health behaviour across diverse groups of men (O’Brien et al., 2005). Men are also hesitant to seek medical help for fear of wasting the physician’s time or being seen as overly concerned with their health (Connell, 2005). Showing vulnerability in the company of another man, for example a male physician, can be particularly damaging to one’s masculinity (Oliffe & Thorne, 2007). In fact, men may trivialize significant health issues for fear of their masculinity being challenged (O’Brien et al., 2005). Therefore, we need to better understand how cultures dictate everyday interactions and social and institutional structures help to sustain and reproduce men’s health risks (Courtenay, 2000).

Women in the lives of men - spouses, girlfriends, mothers or sisters - can play a significant role in men’s health help-seeking. Whether it be a man with an acute injury or chronic illness, women often influence men’s help-seeking (O’Brien et al., 2005). For example, women may motivate men to see a physician (O’Brien et al.), provide emotional support during difficult times (Dieperink et al., 2013), assist in remembering important illness information (Oliffe & Thorne, 2007), or assist with general health preserving activities (e.g., balanced diet, exercise) (Mróz, Chapman, Oliffe, & Bottorff, 2011).

Women are known to engage in health preserving behaviours, including regular exercise, losing weight, and taking vitamins (Courtenay, 2000). Men engaging in such behaviours can be perceived as weak or ‘sissy’, a subordinate masculine position. Therefore, some men distance themselves from health preserving activities for fear of being seen as feminine. For example, men may be hesitant to put on sunscreen as putting cream on one’s body is considered a
feminine precaution. Yet, not applying sunscreen on a regular basis puts men at a higher risk for skin cancer (Courtenay, 2000). Similarly, showing off how much a man can eat and not being concerned about weight can be used to demonstrate a particular masculine performativity, in clear opposition to femininities. However, these practices put men at higher risk for health problems (Courtenay, 2000).

Enacting masculinities also depends on the resources one has to access power depending on social context and situational factors, including “age, ethnicity, social class, and sexuality” (Courtenay, 2000, p. 1390), or financial resources (O’Brien et al., 2005). For example, to demonstrate physical toughness an upper class man may choose to participate in competitive sports. However, a poor man may have limited access to competitive sports due to lack of money, so he may have to resort to street fighting to prove his masculinity (Courtenay, 2000). The belief that “the most powerful men among men are those for whom health and safety are irrelevant…” (Courtenay, 2000, p. 1389) can drive some men with limited means toward risky practices. Violence, smoking, alcohol use, or crime can be used to demonstrate one’s masculinity and may constitute protest masculinity; however, these actions can significantly compromise men’s health.

There are some men who engage in health preserving activities and don’t hesitate to seek help for health related issues. O’Brien et al. (2005) suggest a hierarchy of threats to explain why for some men it is acceptable to engage in activities that are contrary to common masculine practices. For example, sexual performance and being a good provider (by holding down a job) are considered important masculine markers; men with prostate or testicular cancer were willing to seek medical help in order to preserve sexual functioning and continue to support their family (O’Brien et al., 2005). Additionally, firefighters were willing to seek medical help and engage in
health promoting activities because it enabled them to continue to do their job, a role associated with masculine attributes (O'Brien et al., 2005).

Courtenay (2000) discusses in detail how the institutions in society help sustain the health risks associated with masculinity by setting different expectations for men and women. It is suggested that institutions including the government, corporations, industries, academia, media, and the health care system provide structures that reinforce hegemonic masculinities. In the workforce, dangerous work is done predominantly by men, be it in construction, forestry, mining, transportation, etc. These structures expose men to higher risks for injuries, illness, and death. According to Courtenay (2000) even if not doing a dangerous job, the expectations on men to work long hours, deny stress, and not complain about work conditions forces them to take on practices that harm their health in the long run.

Institutional structures are not the only agency reinforcing these masculine expectations; by complying with the expectation in the workplace, men also sustain these masculine ideals (Courtenay, 2000). Additionally, the health care system, for example, pays little explicit attention to men’s health issues and there are fewer programs and resources available to address men’s health issues (Courtenay, 2000; Oliffe & Thorne, 2007).

2.6 Masculinity and Men’s Health Issues

2.6.1 Masculinity and Heart Disease

Heart disease has historically been considered to be a predominantly male disease (Riska, 2006), yet White and Johnson (2000) suggest men delay seeking medical help despite experiencing symptoms that are associated with heart disease. Although delaying seeking help may be partially explained by a lack of knowledge about the significance of chest pain and its association to heart disease, a social and emotional component is also suggested (Robertson et
al., 2010; White & Johnson, 2000). To present themselves as masculine, men constantly try to fulfill the expectations that society places on them: being strong, productive, and fulfilling their roles. However, being sick threatens their ability to fulfill an array of masculine ideals which in turn leaves disabled men vulnerable (White & Johnson, 2000).

White and Johnson (2000) suggested that facing this vulnerability, men tend to deal with the discomforts in one of two ways, rationalization or denial, both of which lead to delaying seeking medical help. Rationalization is the process when men try to explain the discomfort as an insignificant problem (indigestion, stress, tiredness, or normal part of aging) and therefore put up with the discomfort instead of seeking medical help. The second strategy includes men recognizing that their symptoms could be indicative of a serious medical problem but vulnerabilities about having an illness leads them to deny symptoms for fear of the consequences (i.e., not being able to fulfill their roles as expected by society, or being seen as weak) resulting in delayed help-seeking. The degree of the discomfort and debilitation needed to be substantial in order to justify men seeking help for it (White & Johnson, 2000).

White and Johnson (2000) concluded that “the process of male socialization” (or the socially created concept of masculinity) makes it difficult for men to seek help and adopt health preserving behaviours. Similarly, studying the role of masculinity in a man’s decision to seek medical help for chest pain among white (British) and South Asian men (Indian and Pakistani), Galdas et al. (2007) found a difference in how men conceptualized the meaning of seeking medical help that can be associated with the cultural conceptualization of masculinity. Most white men were reluctant to seek medical help or discuss their symptoms with others for fear of being seen as weak. Those white men that previously obtained knowledge about heart disease and its symptoms didn’t hesitate to seek medical help; however, they still emphasized that their
rationale to seek help was not because they couldn’t tolerate the pain, but because they knew that their condition was serious. In contrast, South Asian men who lacked knowledge about the symptoms of heart disease discussed their symptoms with family members or sought medical help quickly resulting in receiving treatment much sooner. Also, South Asian men who were knowledgeable about symptoms of heart disease sought medical help immediately. Discussing their symptoms with others was not perceived as a threat to their masculinity because wisdom, education, responsibility for their family and their health were considered important male attributes (Galdas et al., 2007), a perception of masculinity that is different from the Western ideal of masculinity.

2.6.2 Masculinity and Prostate Cancer

A man’s masculine identity can be altered in many ways due to diagnosis and/or treatment of prostate cancer (Cecil et al., 2010; Oliffe & Thorne, 2007). Sexual prowess is synonymous with masculinity and prostate cancer and its treatments can cause erectile dysfunction, decreased libido, and urinary incontinence (Connell, 2005; Oliffe, 2005). Additionally, men with prostate cancer often experience treatment related body changes that are associated with feminine bodies including weight gain, gynecomastia, and hot flashes (Cecil et al., 2010; Dieperink et al., 2013). While men with advanced age may have an easier time accepting such changes, diagnoses are increasing among men in their 40s and 50s by virtue of prostate cancer screening (Dieperink et al., 2013; Oliffe, 2005). Men with a strong sense of masculinity from other sources (i.e., work, wealth, power) may be able to minimize the threat to their masculinity caused by the sexuality side effects of prostate cancer (Oliffe, 2005).

Being able to work and provide for the family gives many men purpose, identity and a protector role within the family. Most men with prostate cancer were not able to work, play
sports or fulfill household responsibilities resulting in experiencing financial hardships and placing additional burden onto spouses (e.g., restrained finances, additional responsibilities) (Cecil et al., 2010). Those men who had a strong sense of being a provider and protector within the family found it difficult to accept the fact that now they needed help and support from others (Cecil et al., 2010). Facing these challenges, most men felt the need to downplay or hide their struggles from family but also from other men who didn’t experience the same challenges (Cecil et al., 2010; Dieperink et al., 2013; Staples, 2009). However some found it useful to seek out professional counseling that made them feel less like a sissy (Cecil et al., 2010).

### 2.6.3 Masculinity and Depression

For men, admitting to depression and seeking help for it can be particularly challenging as it threatens a range of masculine ideals (Oliffe & Phillips, 2008), for example, being strong, tough and independent. Additionally, depression has been characterized as a predominantly female disorder (Emslie et al., 2006; Oliffe & Phillips, 2008), thus it is also seen as incompatible with being a man (Emslie et al., 2006). Further, stigma is strongly attached to male depression (Coen, Oliffe, Johnson, & Kelly, 2013) particularly where traditional masculine values predominate. For example, in blue collar communities men are expected to embody self-control, strength, responsibility, and being a good protector and provider. When men attempt to share details about depressive symptoms with other men, they risk ridicule, being known as weak (for losing control over emotions), and subordinate within masculine hierarchies (Coen et al., 2013). Therefore, denying depression and avoiding talking about emotions is one way to avoid being assigned to a lower status of masculinity (Emslie et al., 2006) resulting in men trying to self-manage depression (Oliffe & Phillips, 2008).
Brownhill, Wilhelm, Barclay, and Schmied (2005) suggest that because men don’t deal with emotions, their stress builds up and puts them at higher risk for suicide. This may explain why statistics consistently show lower male depression rates but higher rates of suicide compared to women (Canetto & Cleary, 2012; Oliffe & Phillips, 2008). Men who experience depression are likely to present as irritable and act out with rage and anger attacks indicating a different experience of depression than women (Oliffe & Phillips, 2008).

It appears that men who hold strong beliefs of and adopt behaviours consistent with hegemonic masculinity have greater health risks (Courtenay, 2003). Consistent with the finding by Courtenay (2003), “men who aligned to traditional masculinity were more likely to experience depression and less likely to seek help” (Oliffe & Phillips, 2008, p. 195). However, masculinity is dynamic and multi-faceted depending on the context (Canetto & Cleary, 2012). Modifying masculinity can be utilized to preserve health through effectively coping with depression (Coen et al., 2013). *Legitimizing alternate masculinities* by recognizing the need for work-life balance, obtaining self-fulfillment through work and family was a strategy successfully used to cope with depression (Coen et al., 2013). Gender relations within intimate relationships can also be negotiated to co-construct alternate masculinities. Selecting and portraying some idealized masculine behaviours (e.g., building a house, be a mechanic, sexual prowess) can enable men to embody masculinity (Coen et al., 2013). Some men resort to the concept of *protest masculinity* (Connell, 2005) by using alcohol, smoking, drugs and other addictions to try to cope with depression, but this typically lead to a downward spiral into deeper depression (Oliffe, Kelly, Bottorff, Johnson, & Wong, 2011).
2.6.4 Masculinity and Hemophilia

While conducting a qualitative study on men’s experience of hemophilia in New Zealand, it became apparent for Park (2000) that the socially constructed gender order, in essence hegemonic masculinities, played a significant role in men’s experience of hemophilia, developing their identity as a man, and their adherence to best practices in hemophilia management. For example, in New Zealand, men playing rugby is a very strong masculine tradition. Playing rugby embodies ideal masculine attributes, including strength, courage, honour, and discipline, essentially embodying hegemonic masculinity. Being able to play rugby is a key part of developing a male identity especially in adolescence when other sources of masculinity are not available yet (Park, 2000). Indeed, not being able to play rugby caused major distress for boys and men with hemophilia and even their fathers (Park, 2000).

Although some men sought out non-contact sports as alternative activities (e.g., soccer) or engaged in providing administrative support in rugby clubs, for most, these alternative activities were a poor substitute for meeting masculine ideals. For others, the overwhelming dominance of rugby as a national sport served as a constant reminder of their physical limitations (and perceived weakness). For many, the immediate reward of playing rugby (fitting in, being a man) was much stronger than the fear of physical disability (Park, 2000). In addition, the male attribute of stoicism caused delays in treating injuries and contributed to the development of long term joint damage (Park, 2000).

2.7 Summary

Hemophilia is an X-chromosome linked chronic coagulation disorder that is predominantly symptomatic in males and causes internal bleeding episodes (World Federation of Hemophilia, 2013). Males with severe hemophilia experience frequent joint bleeds that result in
irreversible joint damage, disability, lower quality of life, and historically shorter life expectancy (Aznar et al., 2012; Manco-Johnson et al., 2007; Tagariello et al., 2009; Zappa et al., 2012). Prophylaxis is the most effective treatment to prevent joint bleeds and disability and it is used among most boys with severe hemophilia (Aznar et al., 2012; Zappa et al., 2012). However, adherence to prophylaxis significantly drops among adolescent and young adult males and decreases with age while the same age cohort also experiences the highest incidence of bleeding episodes (Fischer et al., 2001; Zappa et al., 2012). Furthermore, even minor or few bleeds can lead to irreversible joint damage, and sustained interruptions in prophylaxis may have long-term consequences (Manco-Johnson et al., 2007; Roosendaal et al., 2008). It is not well understood why prophylaxis is not used by most men with severe hemophilia despite the long-term benefits. Lack of knowledge about the benefits of treatment and long term consequences of not treating is commonly suggested as reasons for lack of adherence but seem to fall short of fully explaining it (Llewellyn et al., 2003; Remor, 2011; Zappa et al., 2012). Research in other men’s health issues suggests that the socially constructed ideal of masculinity is a context that influences men’s health related behaviour. With that in mind, the purpose of this study was to better understand men’s experience of hemophilia in all aspects of their lives and to explore the connections between masculinities and hemophilia across age groups.
Chapter 3: Methods

3.1 Introduction

In this chapter the study design, methodology, theoretical lens, sampling plan, inclusion criteria, recruitment of participants, data collection and analysis, ethical considerations, guiding questions for the focus group, a plan for overcoming possible difficulties, limitations, and the timeline for this research project are presented.

3.2 Research Purpose & Questions

The purpose of the study was to explore men’s experience of hemophilia and the connections between masculinities and men’s experience of hemophilia across varying age groups. There were two research questions:

Research Question #1
How is severe hemophilia experienced among men of varying age groups?

Research Question #2
What are the connections between masculinities and men’s experiences of severe hemophilia across varying age groups?

3.3 Study Design

The study used focus groups to obtain data from men who have severe or moderate hemophilia. Data were analyzed using two qualitative research methodologies. First, a qualitative description research method was used to describe men’s experiences of hemophilia, thereby answering research question number one. Qualitative description is useful for in-depth exploration of complex cultural and contextual health issues, experiences, events, or processes. This method provides a comprehensive summary of the data using everyday language that the
study participants can understand and confirm (Sullivan-Bolyai, Bova, & Harper, 2005) ensuring descriptive and interpretive validity. By avoiding interpreting the data through the lens of the researcher, modification to the data can be minimized. Other qualitative research methodologies (e.g., phenomenology, ethnography, grounded theory) tend to use high level interpretation (e.g., theory development) but these methods risk modifying the final interpretation away from the accounted experience of the participants (Sullivan-Bolyai et al., 2005).

In this study, the participants’ experiences were categorized with a view to describing their principal features. Direct quotes of the participants were used to demonstrate the origins of the categories developed. This represents an appropriate research method for answering research question number one as it only describes a phenomenon without explaining relationships.

To answer research question number two, a constructivist grounded theory research methodology was chosen. Charmaz (2006) described the research process using grounded theory, which allows the researcher to gain an analytical sense of how participants interpret their experiences. Charmaz (2006) suggests that grounded theory is a linear and systematic procedure consisting of nine steps: identifying the research problem, initial coding and further data collection, line-by-line coding, focused coding, writing initial memos and assigning codes to tentative categories, refining conceptual categories, theoretical sampling and collecting additional data, sorting and integrating memos, and lastly diagramming concepts. Constructivist grounded theory uses inductive and deductive thinking to generate concepts, themes and theory, but Charmaz (2006) also emphasizes, that the developed theory is an interpretive portrayal of the phenomenon as researchers inadvertently bring their perspectives and past experiences to the research process. However, this method is still well-suited to allowing the data to give rise to concepts with the least amount of preconceived notions or hypothesis from the researcher.
3.3.1 Theoretical Perspective

The theoretical perspective used for this study was Connell’s *Masculinities (1995, 2005)* which is a framework that permits evaluation of connections between gender and men’s health and it posits that diverse masculine behaviours influence men’s health and illness practices and therefore health outcomes. Connell identifies the following stratifications within masculinities: i) hegemonic masculinity, ii) complicit masculinity, iii) subordinate masculinity, iv) marginalized masculinity, and v) protest masculinity. *Hegemonic masculinity* is a socially constructed ideal form of masculinity whereby men have dominance or power over women and other men. In other words, *hegemonic masculinity* represents the culturally and socially determined behaviours and ideals that claim male dominance. It is typically represented only by a small number of men that produce and reproduce these behaviours and ideals. Different cultures may give rise to a different set of attributes of hegemonic masculinity (Galdas et al., 2007); however, for the purpose of this study, Western societies’ conceptualization of hegemonic masculinity was applied and its attributes include autonomy, control, stoicism, self-reliance, and toughness. The term *complicit masculinity* is used to describe that, although only a very few men are in dominant positions within society, many men are complicit in sustaining hegemonic masculinity and they benefit from the inequalities within society by being dominant over women and subordinating or marginalizing other men. *Subordinate masculinity* is a less powerful form of masculinity than hegemonic or complicit masculinity visible through lower socio-economic conditions, or being stigmatized or excluded from the mainstream society. Additionally, *marginalized masculinity* refers to men whose full participation is limited due to material and structural constraints, such as race (i.e., being a visible minority) (Connell, 2005).
Lastly, Connell (2005) identified protest masculinity that occurs when men with marginalized masculinities overemphasize toughness and risk taking and engage in self-destructive behaviour in order to obtain approval of their “masculinity” from society. Demonstrating dominant masculine ideals of independence, self-reliance, strength, and toughness can garner power for men. However, such behaviours can put men at a higher risk for health related problems and a shorter life expectancy. This framework was used to guide questions developed for focus group discussions and as a perspective to analyze the data.

3.4 Sampling

3.4.1 Sample Population

All participants were registered at the British Columbia Adult Hemophilia Program (hemophilia clinic), run by a large tertiary care center in Western Canada and reported having been seen in the hemophilia clinic within the last year which may suggest that participants represented an engaged group. Of the total of 11 adult male participants (19 years of age or older), three were in the 19 to 24 years old, four were in the 25 to 39 years old, and four participants were in the 40 years or older age stratified sub-groups. All of the participants had severe hemophilia A, except three, who had moderately severe (2% or less factor levels) hemophilia A. Throughout this study, the term ‘severe hemophilia’ is used to refer to the participants in this study with less than 2% factor levels, therefore this includes men with severe and moderately severe hemophilia. Additionally, only one participant reported history of inhibitors.

The participants were from diverse ethnic backgrounds: Canadian (Caucasian or mix of Caucasian and other ethnic ancestry, n=6), Asian (Chinese, n=2), South-East Asian (East Indian, n=1), Middle-Eastern (Iranian, n=1) and Caucasian (Serbian, n=1) ancestry. Of those who
immigrated to Canada (n=5), two immigrated recently (less than two years) and three over 10 years prior to the study. The diversity of participants is reflective of the diversity in the community in this west-coast urban setting (Vancouver and Lower Mainland of British Columbia) that includes a high proportion of immigrants with diverse ethnic backgrounds.

Participants had access to a public health care system in which the goal is to provide universal access for all. Therefore, it is important to note that participants had access to state of the art care without direct cost to them.

Five participants were single (all 30 years old or younger) and six were in an intimate relationship (i.e., married, living with partner). Four participants had a Bachelor or higher education and seven had a diploma, certificate or some college education. Men reported their employment status as working (full or part-time, n=7), studying (n=3) and self-employed (n=1). Joints being damaged from hemophilia included ankles (most frequently reported), knees, hips, shoulders, elbows, and wrists.

Many participants (6 out of 11) had used on-demand treatment in the past and three men had received intermittent prophylaxis in childhood and/or their teenage years. Two participants in the 40 years or older sub-group had acquired HIV infections through prophylactic factor use in the 1980s and had since switched to on-demand treatment. Additionally, all the participants in the 40 years or older and some (n=2) in the 25 to 39 years old sub-group acquired Hepatitis C infections. Eight participants had started using prophylactic treatment in the last few years, though the prophylactic regime varied from daily, three times a week, every third day, and 1-2 times a week. The study was approved by the local research ethics board prior to recruitment and data collection.
3.4.2 Inclusion and Exclusion Criteria

The inclusion criteria were: (a) sex (male), (b) age (19 or older), (c) history of severe or moderately severe hemophilia A or B (clotting factor VIII or IX at 2% or less of normal), (d) enrollment in registration with the Adult Hemophilia Program and (e) willingness and ability to participate in the focus groups.

Excluded from the sample were all those individuals who failed to meet the above criteria. Thus, for example, those who had no interest in participating in the study, or who were unable to attend any of the focus groups or participate in the discussion due to language barriers were not included.

3.4.3 Recruitment Methods

Individuals registered with the Adult Hemophilia Program who met the sex and age criteria were contacted by the hemophilia clinic staff and provided with information pertaining to the study. To maximize recruitment, potential candidates were contacted using several strategies: an information letter and a written invitation to participate in the focus groups were provided during a clinic visit or the same information was provided via emails or mail-outs to those who were not due for a clinic visit.

Individuals interested in participating in the study were provided with a consent form. The consent form provided specifics about the study, including the purpose, format, risks and benefits, procedures for maintaining confidentiality and contact information. All participants’ questions were answered to their full satisfaction prior obtaining informed consent. Potential candidates were provided with a schedule listing several dates for focus group meetings. The project coordinator scheduled the time and location based on the availability of the participants.
and research staff. As a token of appreciation, at the end of the focus group meeting, participants received a $50 gift certificate and were reimbursed for parking expenses.

3.5 Procedures and Data Collection

A qualitative study design is well-suited for generating data relating to the diverse experiences, perspectives, ideas and attitudes of individuals with a history of hemophilia (Polit & Beck, 2012). A semi-structured focus group interview format was selected owing to its efficacy to collect rich information. As per Krueger and Casey (2009), this format is highly efficient for conducting in-depth discussions that reflect both individual and group viewpoints; it is particularly useful, moreover, for collecting data on how perspectives, attitudes, and ideas evolve within a specific social setting. Each focus group was led by two male researchers, the purpose being to build rapport between the exclusively male subjects and researchers, thereby enhancing the richness and accuracy of the data, which could conceivably be jeopardized if females were leading the focus group. Additionally, the researchers were specifically not hemophilia care providers so that participants might speak more freely without fear of influencing their care experience. Lastly, a demographic form was used to capture the background information of participants including their age, employment and marital status, education level, type of hemophilia, inhibitor history, type of current treatment, bleeding frequency, and history of prophylaxis use (See Appendix A).

3.5.1 Focus Group Procedures

Participants were assigned to one of three age stratified focus groups: a 19 to 25-year-old group, a 26 to 39-year-old group, and a 40-year or older group. This stratification was essential as some participants may have had difficulty relating to the experiences of those in younger or older age groups. We anticipated that participants would be more likely to share their
experiences within a group consisting of individuals they perceive to be similar to themselves, and less likely to share their experiences within a group that is diverse in terms of the life experiences of the members. Stratification of participants based on age also allowed us to compare and contrast the experiences of men with hemophilia based on their trajectory with the disorder.

Focus group discussions were semi-structured and lasted approximately 90 minutes. At the beginning of each focus group, the researchers introduced themselves and provided a short overview of their professional backgrounds and their roles in and involvement with the research project. The researchers also reviewed the purpose of the study, answered questions, confirmed that informed consent was obtained, and reiterated the principles of strict confidentiality procedures for the research project. Each session was audio-recorded and a speaker’s log was kept to facilitate accurate data transcription. The researchers thanked the participants for offering up their time and advised them that they were free to withdraw from the study at any time. Next, the researchers informed the participants that their role was to ensure that each one of them had an opportunity to share his thoughts and that the discussion was in no way intended to be an exercise in consensus building, but rather diverse perspectives were welcome. To ensure accuracy of the transcribed data, participants were asked to speak clearly and one at a time. At the end of the session, all participants were thanked for their contribution to the research project.

3.5.2 List of Questions

A list of questions was prepared, the purpose of which was to guide and facilitate a semi-structured discussion on how hemophilia had affected not only the participants’ physical health but every facet of their lives, including intimate relationships, social relations, career prospects
and development, physical activities, life goals and aspirations, and their overall sense of “male” identity (see Appendix B). The list of questions was revised after the first focus group; first, the sequence of questions was changed in order to initially focus the discussion on the technical aspects of their experiences with hemophilia and ease the transition into more sensitive topics, including relationships, stigma, life goals, and challenges. Then, the wording of some of the questions was changed to reflect layman’s terms and ensure open-ended questions were used. Lastly, additional questions were added to the list to explore important discussion points that emerged from previous discussions. All subsequent focus groups used the second format of the list of questions (See Appendix B for the list of initial and revised guiding questions).

3.6 Data Analysis

Each of the focus group discussions was audio recorded. Audio recordings were transcribed by trained and experienced personnel. One researcher verified the transcription against the audio recording for accuracy (LK). The data collected were analyzed for emerging themes and concepts that describe the participants’ experiences of hemophilia for the purpose of answering research question number one. Data collected on the demographic form was used to report the characteristics of the representative sample in the study. Additionally, connections were explored between men’s experiences of hemophilia and the background information collected on the demographic form (i.e., disclosure and prophylaxis use).

The suggested nine steps by Charmaz (2006) for conducting constructivist grounded theory method were modified to answer research question number two. During this study, field notes were taken and preliminary thematic development was started during the data collection phase which guided the modification of research questions as described earlier. However, detailed analysis of the data commenced only after all data were collected. This strategy
minimized the introduction of preconceived notions during all focus groups in the hopes of collecting unbiased data. In addition, line-by-line coding was replaced with incident-to-incident coding that was better suited for comparison of men’s experiences of hemophilia across age groups. While being immersed in the collected data and through coding and writing memos, the writer was able to gradually develop concepts, categories and themes. Constant comparative method was maintained throughout the iterative data analysis process. Data analysis was guided by researchers who are experienced in Charmaz (2006) grounded theory process. Major themes and theoretical categories were also reviewed by health professionals with clinical expertise. Several memos and drafts of the findings were prepared prior to finalizing the findings.

3.7 Limitations

The diverse backgrounds of study participants reflects the community of a West-coast urban Canadian setting and likely contributed to the sharing of diverse experiences, but findings may not be transferable to other communities that are less diverse or that are rural. The relatively small sample size (eleven participants in three age stratified sub-groups) of men with severe and moderately severe hemophilia A only may limit the transferability of the findings to men with hemophilia B and mild or moderate types. However, the findings may be of use in developing a theory of masculinity in hemophilia or in guiding future research focusing on hemophilia management.

The researchers conducting the focus group discussions had limited knowledge about hemophilia and may have missed clarifying clinically relevant points which may be perceived as a limitation. However, the researchers’ lack of clinical knowledge provided an opportunity for the study participants to take on an expert role and ‘teach’ the researchers about their disorder. The establishment of such dynamics during the discussions was a strength to the study because it
facilitated the sharing of men’s experiences and their subjective meanings and enabled researchers to derive findings without preconceived perceptions (Oliffe & Mróz, 2005).

The focus group methodology used in this study is an effective way to obtain diverse viewpoints. However, it is not certain that all participants in this study shared authentic personal experiences and perspectives, for any number of reasons: some participants may have tried to “please” others (e.g., the researchers or other participants), and others may have felt compelled to conform to the consensus view. Thus, while focus group studies are considered to be efficient and accurate at obtaining qualitative data, we can’t rule out the possibility that at least some important perspectives went unshared and that others, though shared, were modified to “fit” the dominant view point.

The voluntary participation of the study may have resulted in biased representation of experiences; however, the diverse experiences included less than perfect hemophilia management practices indicate a balanced representation of experiences. Lastly, the study design is particularly suitable for identification of new central concepts and connections in hemophilia experiences and management practices, but is insufficient for exploring cause and effect associations between those concepts.
Chapter 4: Results

The purpose of the study was to explore men’s experiences of severe hemophilia and the connections between masculinities and men’s experiences of hemophilia across varying age groups. The two major thematic findings arising are supported by excerpts from the data and connections to masculinities are discussed throughout.

4.1 Men’s Experiences of Severe Hemophilia

4.1.1 “Learn Your Body” – Navigating the Limitations of One’s Body

Participants spoke about how, over time, they learned about their body in the context of recognizing the symptoms of a bleed, the activities likely to lead to a bleed, and treatment methods for stopping or preventing a bleed. Over time men became experts in managing hemophilia, but men recognized that in their younger years they were learning how to manage their disease.

4.1.1.1 Know Your Pain

In line with the wider men’s health literature suggesting pain is the symptom most often triaged by men in deciding to seek medical help, the participants in the current study highlighted how pain – as a cardinal sign of a bleed – was the catalyst for using factor and/or seeking professional help. Indeed none of the men referred to other signs of bleeding, such as a joint becoming tight or hot and bubbling or tingling inside a joint. A man from the 25 to 39 year old group explained the warning signs: “…I found before bleeds, I feel some pain …I have some pain and when I feel that pain I infuse.” In this regard pain played an important role in notifying men that there was a bleed that needed to be attended to, as a man from the 25 to 39 years old group eloquently quipped: “Pain…let’s you know, you don’t want to know, but need to know.”
A man in the 19 to 24 years old group acknowledged that diversity can exist among men’s bleeds and identified that bleeds need to be treated in a timely manner: “Because my body reacts very quickly if I get injured. It reacts very quickly. I have to take an injection within [a] half an hour. So, … every hemophilia victim’s body acts differently - not similar at all.” There was consensus that pain had to be attended to because if it was left untreated it would not only increase in intensity it would likely result in residual joint damage - and perhaps immobility and chronic pain. If one experienced any hint of a bleed, the best course of action was to treat and treat early. For example, participants in the 19 to 24 years old group confirmed that any inkling of a bleed should be treated prior to sleeping:

Because I would go to bed, like my ankle would feel a bit off, but I’d say, ‘oh it will be better in the morning, I’m not going to be sitting on it or sleeping on it, or anything like that’. But in the morning, it’s just terrible and you can’t walk, but it’s that, that was when I was younger. I didn’t understand it as much.

Experiencing joint bleeds and pain enabled men to learn what activities were likely to result in a bleed and how their bodies reacted. Walking or running on a hard surface (i.e., concrete) was more likely to result in a bleed compared to doing the same activity on a soft surface (i.e., grass). Even being overwhelmed with other activities such as work, studying or family responsibilities, can result in increased number of injuries, as a man in the 19 to 24 years old group stated: “If I’m busy [with studying or work], I get injur[ed] very quickly.”

In addition, some participants explained how they learned to differentiate chronic arthritic pain from acute pain resulting from an acute bleed. A participant in the 19 to 24 years old group explained that if a sharp joint pain was relieved with warming up the joint then it is consistent
with chronic arthritic pain. Pain due to an active bleed will subside if treated, but chronic arthritic pain will not. Learning to differentiate between types of pain had taught him to identify the correct course of action.

In terms of contrasts across the sub-groups by age, it appeared that the greater the exposure to bleeds and experience with management strategies, the better they became at effective treatment and/or prevention of bleeds. Men in the 19 to 24 years old group were more likely to share experiences of their management strategies in the focus group discussion. The sharing of experiences was beneficial to validate one’s experience and also to learn from others. In contrast, men in the 40 years or older group were confident and steadfast in their experience and management strategies. This age difference suggests that while men in the 19 to 24 years old group were still learning to manage hemophilia, men in the 40 years or older group became experts in managing their disorder.

Within the focus group interviews the individuality of participants was evident especially in terms of their experience of bleeds and treatment regimes. So while other men with hemophilia might be a reference point in some regard, their individual treatment regimens tended to vary greatly. This informed an interesting dynamic whereby the participants often aligned with one another as sharing a disease but their illness management strategies were individualized for the most part and drew on strength-based masculine ideals including autonomy, problem-solving and self-reliance. As a man in the 40 years or older group clarified that knowing his pain and body was the gateway to effective treatment:

…to me, it’s, it’s very, um, case specific right? It’s whatever the bleed is, it’s – have you noticed a pattern? How much pain are you in? You know what I mean? Like, if it’s a joint, I know how that joint is typically gonna respond or react in advance to a particular
activity. If it’s muscular, sometimes I - I’m working out and I have a muscle that gets kind of uppity and it’s worked, you know, it seems to be a problem for a while, then I’ll treat a little more aggressively over a longer period of time. It’s case by case.

Men reported taking hemophilia management very seriously, but, for the most part, they reported being less aggressive with other health related issues: “anything that’s not injury related to hemophilia, it’s no big deal at all.” For example, stomach ache, headache, cold or flu “would go away” on their own. This attitude is consistent with men’s health help seeking practices in other men’s health issues, in which symptoms need to be significantly debilitating for men to consider seeking medical help.

4.1.1.2 Navigating Treatment

Finding the right treatment regime that matches the individual’s level and frequency of physical activity involved a certain amount of trial and error even when supported by expert hemophilia clinicians. The most common prophylactic regimes included infusing two or three times a week typically during the week days, which didn’t always fit with an individual’s activity level. A man in the 19 to 24 years old group experienced breakthrough bleeds: “I’ve tried different regimes, like two times a week, three times a week, but if I was infusing on the day of one sport and not on the other, I found that I would have breakthrough bleeds.” Similarly, another man in the 25 to 39 years old group found infusing during the week resulted in “struggling for two extra days” on the weekends and the tendency “to bleed more after that.” Adopting a regularly scheduled regime that was not associated with dividing doses across a week resulted in fewer bleeding episodes for one participant: “I follow three days instead…[I] get bleeds like once a year maybe”.

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Participants expressed how today’s factor concentrates make self-infusion possible, a significant advancement in care as it eliminates the time needed to receive treatment in a hospital setting and also increases their freedom. Although some reported disliking self-infusions: “Yeah, I think it’s [a]pain, I think it’s horrible. I hate it” stated a man in the 25 to 39 years old group. However, it is perceived to be a better alternative than having to go to the hospital for every treatment: “…in the old country, I used to go to the clinic, but it’s much easier to do it yourself. I mean it takes time to learn, but once you start doing it then it’s easy, and it also saves time.”

Maintaining venous access for treatment was a concern across all age groups, but to a greater extent among the 40 years or older group. Some men, particularly among the 19 to 24 years old group, described how efficient they had become with self-infusion, how easy it is to find the vein, but were aware that it may change with age. Others strategically chose treatment regimen to somewhat minimize the frequency of infusions to preserve their venous access. A participant in the 25 to 39 years old group described limiting the post-bleed follow-up infusions to once a day instead of twice daily: “…when I treat, I don’t actually do the full twice a day thing, I do once. So I follow up once every day after. I’ve tried that because I don’t like to blow up all my veins, because I only used two veins for the last 20 years or so.” However, participants in the 40 years or over group reported that the self-infusions are starting to become more of an ordeal due to poor vein access and deteriorating eye sight:

I find, especially as I am a little bit older, I’m finding the venipuncture a little more challenging than I ever did. It was never even [a challenge], I mean you know, blindfolded with my feet, you know, upside down, no problem. But in the last couple of
years, the eye sight’s not as good, the veins aren’t as grabby of the needle, so, you know, it’s a little bit more of an ordeal, I guess.

Self-infusion also makes prophylactic treatment possible, a further advancement in care by preventing bleeding episodes. Prophylaxis wouldn’t be feasible if infusion were limited to hospital or clinic settings because of the time and inconvenience involved. Most men emphasized the advantage of prophylaxis in preventing joint bleeds. Yet, adhering to prophylactic treatment can be challenging for several reasons, including the frequency of infusions and long term venous access as described above. Additionally, a participant in the 19 to 24 years old group stated that he doesn’t use the factor prophylactically because of its short half life (less than 24 hours) as he experienced breakthrough bleeds shortly after prophylaxis: “Yeah, because sometime I got [an] injury right after the injection after 18 hours, I noticed,” which makes the factor more suitable for stopping a bleed for this person (on-demand) than for prophylactic treatment. Experiencing major injuries may have motivated men to start prophylactic treatment by realizing that they need to take better care of their bodies, as a man in the 25 to 39 years old group with a previous left knee injury described: “I injured the right knee, and that was kind of, oh, that’s when I got really serious about stuff, and I started doing more like prophylaxis.”

Another significant difference among the age groups was that while prophylaxis enabled the younger men with relatively healthy joints to be physically active, it was insufficient for men in the 40 years or older age group with advanced joint damage: “No amount of product is going to make that [damaged joint] better.” Prophylaxis can be perceived as “too late now” by men with advanced joint damage and disability. This attitude is reflected in treatment regimens, as 3
out of 4 participants in the 40 years or older group were using on-demand treatment. Despite these challenges, prophylactic treatment, including daily prophylaxis, has been increasing over the last few years across all age groups. Indeed, a participant in all age groups relied on daily prophylaxis.

The care that Canadian immigrants received in their country of origin was not necessarily comparable. As a man in the 25 to 39 years old sub-group explained that he considered himself doing well compared to others with hemophilia in his country of origin, but not as healthy and fit when compared to Canadian counterparts. Evident here and in other immigrant men’s narratives were the bittersweet benefits of having access to effective treatment albeit having musculoskeletal deficits that may have been preventable. In this context, participants who craved and complied with treatment ran counter to Western masculine ideals that prescribe resistance to health help seeking as the norm. Furthermore, none of the immigrant men used daily prophylaxis which may indicate that uptake of prophylactic treatment and its frequency may be gradual and dependent on previous norms.

A certain level of skepticism about the absolute safety of the current factor products was present across age groups as they were all aware that plasma derived factor concentrates carried viral infections, such as HIV and Hepatitis C in the 1980’s. However, they also recognized that without alternative treatment, they don’t have a choice but to use the products: “we don’t have a choice, so we use it.” Such dependence on factor availability and recommended treatment reflected their vulnerable position within the health care system which they are aware of.

Two of the four participants in the 40 years and older group acquired HIV, while all participants in the same group and some (n=2) in the 25 to 39 years old group acquired Hepatitis C infections from factor use. The spread of infectious diseases through factor products had the
largest impact on the 40 years and older group, which may help explain the strongest skepticism among this group. As a participant in the 40 years and older group eloquently expressed that the system is much safer now, but believes that bad things could still happen:

...I [my] hope is that (clears throat), is that we have a safer system than we did 20 years ago and I have become convinced of late that we do. But that doesn’t mean that [I] would be shocked if tomorrow I got ...[informed that]... I have some funky new bug...Would I be gob smacked? No. Not at all.

Although none of the participants in the 19 to 24 years old group were directly impacted by infectious diseases from factor use, they still carefully assessed each product for the risk of infectious diseases. Their understanding was that most factors are similarly produced and safe, but felt most secure with products they used for a long period. Hence, many would likely not be an early adopter of a new product, as a participant in the 19 to 24 years old group stated: “Just yeah, the only thing I would be worried about is changing to...one of the possibly new products...I wouldn’t be an early adopter.” Skepticism is not necessarily limited to factor products only, but may include a new treatment regime. For example, a man in the 25 to 39 years old group expressed that he understands the advantage of daily prophylaxis, but he is not ready to embrace it yet: “I might actually try that routine once the studies go through...Right now I’m not sure. I’m letting it ride for a bit. Everybody says it’s great though.”

Many men emphasized the need to take responsibility for managing their hemophilia to effectively navigate treatment options, and it may sometimes mean going against the advice of a health professional: “you know enough not to let doctors tell you what to do because they’re not always right”. A man in the 40 years and older group rationalized that he managed to avoid
acquiring HIV by staying on cryoprecipitate (and on-demand treatment) against physician’s recommendations:

When I was younger, when it [factor concentrate products] started to come in and I was at a particular clinic back in [city name], and they were heavily pressuring us to switch to the combined concentrates, my parents had read in a magazine about HIV being transferred in those drugs in the US and they said “no”, and when the doctors said, “well, we’re not going to give you a choice” we switched [clinics]. Then, I stayed on cryo [cryoprecipitate] for another 10 years prior to switching to concentrates and I actually ended up getting Hep C, but [avoided HIV].

4.1.1.3 Managing Risks

In addition to hemophilia treatment, learning also took place around ensuring that the factor was available when needed for treatment without delay and stored properly at locations where they spent most of their time (primarily at home, possibly some at workplace or school). Essentially, all activities needed to be planned for in advance: “I think there’s always risk associated with hemophilia, no matter whether it’s work or travel [or leisure], and I learned to bring with [me] my own factors to travel wherever [I go].” Travelling to other countries brought additional challenges such as consideration of health insurance, availability of product, and location of nearest hemophilia clinic. For air travel, keeping the factor in carry-on luggage (not to risk losing the luggage) and having documentation prepared that justified the need for the factor (for airport security) were practical steps men learned to ensure that they were always self-sufficient with factor products. As one participant quipped in explaining the need for always planning ahead with hemophilia to ensure timely treatment: “you may be on vacation, but you are never on vacation from hemophilia.”
Participants reported that one always has to make decisions on what level of physical activity one can engage in: “You can do everything up to a certain point right?...even the healthiest people can get injured,...but for us, the healing process and everything is more difficult. It’s “up to the individual to take the risk.” The type of physical activity men reported to engage in carries varying degrees of risk for bleeds and included swimming, free diving, bicycling, curling, ultimate frisbee, golfing, cricket, soccer, weight-lifting, snowboarding, walking, running, and even motorcycling. The participants recognized that some of these activities could be considered high risk for bleeds, but they rationalized that the risks could be minimized by infusing prophylactically prior to the activity and being extra cautious. The risk for an injury by engaging in a particular activity was evaluated by participants in the context of what activity one was passionate about and how the risk can be managed, a process that can be rather subjective. For example, commuting by motorcycle on a daily basis without prophylactic treatment was perceived as a low risk activity by a man in the 19 to 24 years old group. Similarly, a participant in the 40 years and older group infused prior to free diving “to ease the fears of free diving instructors and hematologists worldwide,” but he was “not sure that it makes [made] any difference for that activity.” These examples depicted masculine values of individuality, independence and risk taking when deciding what physical activities to participate in. Indeed, men utilized participation in high risk physical activities and disregarding potential risks as a form of demonstrating masculinity. Men in the 19 to 24 years old group were particularly likely to engage in higher risk physical activities.

Participants across all age groups were aware that joint injuries could limit the type and duration of the activity one could engage in, as a man from the 19 to 24 years old age group described it: “well, now that I’ve had those injuries [shoulder injury], they definitely limit my
ability to workout [in the gym]...so I have to work around that to try to buffer the shoulder.”

However, physical limitations increased with advancement of joint damage and age. For example, a man with ankle injuries in the 25 to 39 years old group explained that running and playing soccer is difficult: “I can’t play soccer, I, actually I can but if I play then I have bleedings, so I prefer not to play. So, it makes my activity less compared to other normal people.” Most men in the 40 years and older group “don’t [didn’t] have the kind of lifestyle” that involved sports and were limited to walking only due to the advanced joint damage and muscle wasting. Even playing low intensity sports like golfing was difficult for them due to lack of “balance” and “stability” caused by weak joints.

Participants across all age groups portrayed understanding the benefits of regular exercise to strengthen the joints, at least in theory. However, some participants found that trying to strengthen the muscles through regular exercise can be a “vicious circle” as increased exercise resulted in bleeds that interrupted the exercise regime and negated any physical strength gained. Others found regular exercise beneficial with the right preparation and support. Prophylactic treatment prior to an activity, doing strength exercises without impact (i.e., spinning, cycling, and swimming), use of light weights, and relying on the guidance of personal trainers were effective strategies of preventing bleeds. Men in all age sub-groups offered the advice of doing everything possible to prevent joint injuries early on, particularly ankle injuries because it can be particularly limiting for them. There didn’t seem to be an age related difference in men’s willingness to engage in regular exercise to protect and strengthen their joints; however, previous individual experiences and perceptions were likely to influence individual practices.

Some men considered the use of supportive equipment, such as ankle or knee braces, a way of preventing joint bleeds during physical activities and suggested their use be promoted to
children: “tell every young kid to get ankle braces” at a young age to prevent joint damage as much as possible. Others expressed concerns that their regular use may actually weaken the muscles around a joint and increase the risk of injury. Their perception of the benefit of supportive equipment seemed to be different from their intended use from the clinical viewpoint of providing support to correct gait problems and prevent deterioration of other joints. Additionally, some voiced concern that wearing protective equipment might make one “look like a bionic man” and risk being ridiculed by others indicating that hemophilia can have an emotional or psychological impact on men. In addition to taking responsibility for hemophilia treatment, men needed to ensure factor availability in all circumstances, manage engagement in physical activities and cope with physical limitations caused by joint injury.

4.1.2 “Don’t Let It Hold You Back” - Negotiating One’s Identity in the Context of Hemophilia

Most study participants were limited in the physical activity they were able to engage in. The physical manifestation of the disorder also impacted virtually all aspects of their lives including type of work available for them, social relationships, and roles within family or among friends that challenge their masculine identity. Western societies’ concept of hegemonic masculinity includes physical strength, independence, stoicism, being a good protector and provider, and having sexual prowess. Although all men have to work towards constructing and re-constructing behaviours in relation to hegemonic masculinities, men with severe and moderately severe hemophilia strived for the same values from a subordinate or marginalized position because of the physical manifestation of hemophilia.
4.1.2.1  Coping with Limitations

4.1.2.1.1  Vigilance for Potential Physical Danger and Limitations

Participants across all age groups reported that the risk of injury was virtually always present because of the significantly higher risk for internal bleeding episodes compared to the general population performing the same activity. This circumstance created a constant sense of being vulnerable to physical danger and resulted in constantly assessing for risk of injuries – an increased vigilance to physical danger and becoming subconsciously aware of dangers and limits:

I find myself more conscious of my surroundings, I find myself checking, making sure I know where things are subconsciously and everything. It’s really odd, but I think that’s just one of the things that you acquire as you get older with it [hemophilia].

Another man reported that “it [vigilance] just blends into your whole kind of life” even “with regards to studying” as “[you] start to know what you can do, when you need to ask for help.” Although the heightened spatial awareness was an advantage in some activities, for example motorcycling or playing ultimate frisbee, or learning one’s limits in studying, overall it was a burden that was always present: “I find, I spend a lot more time thinking about physical risk in what you are doing and... just how I thought about things and the amount of thought I gave, or the issues you’d have to go through.”

Regardless of age group, all men related that hemophilia placed limitations on the physical activities they could engage in: “...it affects you in every way because...you can’t do the same things like other, like people who don’t have hemophilia right?...you can’t do everything right? And many times you want to do something and you just can’t.” As their physical
activities are limited, men with severe hemophilia generally lacked the physical strength, a hallmark of socially constructed masculine ideal, and it impacted their perception of being a man. The awareness that they don’t have either the physical strength or the muscle mass needed for a masculine appearance created an inferiority complex, as a man in the 40 years and older group expressed it: “you see all the strong, muscle guys and then you say, oh, you couldn’t even have the weight to do the bike or something. So you got, you got the infer- inferiority complex.” Not having the physical strength to participate in some activities resulted in a self-limiting mentality as a man in the 40 years and older group expressed it: “I think that, you know, that there’s a lot of things that you think, maybe even because of the hemophilia that ‘oh I can’t do that or that’s not for me’ or that’s not available to me” and not even try to be active: “I never think of having to go down to the gym and have a personal trainer, that’s not in my mind at all.”

Attempts at regular exercise without proper preparation and support resulted in bleeding episodes without gains in physical fitness which served to further reinforce inferiority and self-limiting mentality. Hence, some men with severe hemophilia “accept[ed] the fact” that they can’t be physically active. Although some participants, especially in the 19 to 24 years old group with relatively good joints, reported engaging in regular exercise, older men suggested that all men could be more active with the right preparation (i.e., prophylaxis), education (i.e., minimizing impact with strengthening exercises), and viewing physical fitness as a self-improvement process. As a participant in the 40 years and older group put it: “your number one competitor is yourself” and try “not comparing yourself to others so much when it comes to sport and athletics.”
4.1.2.1.2 Social Engagement

The physical limitation experienced by participants across all age groups also impacted their interests and commonalities with others, and hence impacted their social relationships. Some men excluded themselves from social settings where people were physically active, such as running, hiking or playing soccer because of the physical limitations caused by hemophilia, as a man in the 25 to 39 years old group expressed it: “It’s difficult for me to run, so I can’t be in those communities where people are running.” Social relationships also influenced treatment decisions. Men reported that particularly during childhood friends not understanding their disorder indirectly influenced them to delay treatment by encouraging them to ignore a bleed or injury and continue playing. Supportive statements such as “do you need to go take a shot” or “are you sure you want to do this” by friends who understood their disorder encouraged them to look after their bodies. Compared to younger men, the older men stated that they had learned to take responsibility for looking after their disorder regardless of social influence and also establish supportive social relationships.

Men also found it beneficial to connect with others who had hemophilia to exchange experiences and ideas, drawing solace that they were not alone in tending to the challenges accompanying hemophilia. Social events within the local hemophilia community, such as family gatherings, camps and outings were important opportunities for peer support and information sharing as some portrayed the importance of learning from others’ experiences in addition to what health professionals were able to offer in hemophilia management decisions. Although the need for being a part of a tight knit hemophilia community can change over time, it was particularly beneficial during the early years. However, men shared that they found it difficult to reach out to and connect with others with hemophilia through the national hemophilia society.
due to the strict confidentiality rules: “the national organization has a website but it’s not really setup to facilitate connections between” members. Particularly men in the 19 to 24 and 25 to 39 years old group identified the need for more peer support opportunities.

Parents played a dominant role in managing hemophilia in childhood. Men across all age groups recognized that parenting a child with hemophilia can be challenging to try to find the right balance between protecting the child from physical harm without being overprotective. They all resented when parents restricted their activities, but now understood the need for it. Still, they recommended to other parents of children with hemophilia to let the child lead as normal a life as possible and to not scold a child after an injury because they will more likely hide an injury in the future. Being overprotective may work for a while, but there will be a time when parents won’t be able to stop them from participating in ‘riskier’ physical activities.

According to participants in all age groups, the hemophilia clinicians had a significant influence on their development at a young age. Men thought back very positively about the clinic staff that supported them through difficult times:

The nurse coordinators of the programs are, you know, they are like the mother hens, they do a lot of everything. I had some amazing people who I’ve had great relationships with over the years, like people I remember as being like very, very special in my life because they are so instrumental in helping you through tough stuff.

### 4.1.2.1.3 Work and Career Obstacles

Hemophilia limited the type of work that participants were able to do because of the physical limitations the disorder placed on them, as a man in the 40 years and older group expressed it: “[we’ve] got to be careful about the work we choose because being a hemophiliac, you cannot take every kind of work you want.”
All the participants related to the physical limitations hemophilia place on their work options, as a man in the 19 to 24 years old group illustrated it: “I was going to be doing lifeguarding, but of course broke my shoulder. Never been the same since. And then my ankles of course are pretty injured. So, that physicality of hemophilia will always stay with every hemophiliac.” Although men in the 19 to 24 years old group might have been able to carry on with physical labour for a while with the right treatment regimen, they recognized that injuries will likely restrict them relying solely on it and were generally motivated to pursue higher education: “it steers you a little bit towards higher education.” Advanced joint damage limited them to sedentary jobs, particularly men in the 40 years and older age group. However, limitations applied not only to the physical aspects of a job.

Their work and career was further limited because even simple activities that other people take for granted, like traveling or moving to another city for work, was more complicated by having to plan for factor availability, health insurance, air travel security, location of nearest clinic among others, as a man in the 40 years and older group explained:

You have a support system that you already established, and that you feel comfortable in.

I got like four doctors. I mean, I don’t want to get four new doctors, you know. So, professionally, any of those decisions that you have to make, beyond the physical risks, lifestyle wise, become an issue, you know.

In addition, some of the men in the 40 years and older group were unable to plan ahead to maximize work or career potentials because of the shorter life-expectancy with hemophilia prior to factor concentrates and due to HIV infections. Their life goals and aspirations were somewhat different than the typical population in that planning for work and career was not a priority and it
now places them in a disadvantaged position. As a man in the 40 years and older group reflected on his work plans and aspirations:

Knowing that I had that [HIV], and in 1987 that was not good news, it kind of derailed [me] personally, it derailed me for a number of years. I ended up getting involved with, you know, AIDS advocacy, they were inspiring, exciting times, but they didn’t prepare me at all for this beautiful long life that I have ended up having, and led to a break from that continuum which when I finally sat down, kind of 15-20 years later. ‘OK, what am I gonna do with this life?’ It was in some respects, a little bit too late.

Work plays a central role in constructing men’s socially defined masculine identity and many men align to provider and protector roles. The type of job, career and salary oftentimes determines men’s masculine capital and in this context hierarchies emerge based on the work men do. In North America, being a hockey player is idealized, and the courage and self-sacrifices that firefighters, police officers, or soldiers make are also affirmed as the work of honorable, brave men. Study participants across all age groups stated that they understood such celebrated and revered masculine work was not really an option for them. Indeed, men with severe and moderately severe hemophilia might be known by the work they couldn’t do rather than defined by the work choices they could sustain. While younger men were steered toward higher education, older men with limited education were the most disadvantaged in work and career options in this study.

4.1.2.2 Striving for Independence – Taking Control of Treatment

All study participants emphasized being independent with managing hemophilia, an attribute consistent with hegemonic masculinity, or at least they strived to present that image.
One of the participants initially commented that he felt like hemophilia was managed for him by others (by hemophilia clinic staff), but later presented independence in treatment decisions: “I treat when I feel like I need to treat, not when someone else says I should treat.” Although men acknowledged the important support they receive from hemophilia clinic staff in managing hemophilia, they all emphasized that others (i.e., spouse, family) don’t have any input into how they manage hemophilia.

Men portrayed that the pediatric hemophilia clinic instilled a lot of independence and responsibility in them by facilitating learning self-infusion, typically around the age of eleven, but acknowledged that predominantly their parents directed hemophilia management in their childhood. Men in the 19 to 24 years old described their high school years as the time when they experienced a lot of bleeding episodes and joint injuries: “High school was probably the time when my ankles got the worst.” Reflecting on their earlier years, men recognized that while their parents directed hemophilia management in their childhood, high school was the period when they started to become independent in managing hemophilia for the first time and they made mistakes that resulted in irreversible joint damage.

Men partially associated the increase in bleeding episodes with their increased level of physical activities and participating in sports. They also acknowledged that a learning curve took place around hemophilia management: physical limitation and adjusting treatment. In addition, men also associated the high incidence of bleeds and joint damage in high school to their lack of understanding of the long term consequences: “...it was just because I didn’t understand the impact it would have in the future,” and having a sense of being invincible: “I want to have fun now and I’ll worry about it later because there’s this medicine I can take.” Men also reported hiding treatment from classmates by delaying infusion until the end of a sports
game or end of the day for wanting to fit in and not showing vulnerability, another masculine attribute: “you are just like ‘I’m in gym class, I’m going to play and I’m gonna play hard’. And then later that day you are like ‘oh, no, my ankle hurts’.” Other elements that may be at play during this period is their need to prove their masculinity by challenging themselves physically, similar to findings by Park (2000) among New Zealand men with hemophilia needing to play rugby, as a man in the 40 years and older group eloquently explained:

I feel like I fell in with the wrong crowd in high school, and it wasn’t drugs or alcohol, it was, I fell in love with the basketball crowd and I took great pleasure in playing basketball, but that was a poor choice. And, I guess, my advice would be to find other ways to meet those needs that I know I had as a young man, you know, to challenge myself and to play, and to find ways that, other ways to get those needs met

Although mostly the youngest generation spoke of the frequent bleeds in high school, men in all age groups related to that. Men also expressed how much better they became at assessing activities for potential injuries by the end of high school, indicating that learning took place from those mistakes. Knowing one’s pain was the conduit to effective self-management both in terms of treating spontaneous bleeds as well as prompting participants to thoughtfully consider using factor prophylactically. Furthermore, these findings suggest that men with hemophilia engage in physical activities to prove their masculinity and fit in with other men even if it comes at the price of irreversible joint damage, a behaviour consistent with suggestions by Courtenay (2000) that men engage in behaviours that is detrimental to their health for seeking masculine status. The need to construct their masculinity through physical prowess may be
particularly prevalent during the high school years when other sources of masculinity are limited, such as social status, wealth or work.

4.1.2.3 “I am not this” – Pushing Back Against Hemophilia

Although self-infusion and prophylaxis provided a sense of freedom by allowing men to be self-sufficient and participating in more activities to the point that “some days you don’t even think that you have any disease at all,” for most men across all age groups, it was too much at times to think about all the impacts hemophilia has had on their lives: “we have to basically think about it or sometimes, not think about it.” For example, having to stay home to recover after a major injury or seeing other children playing sports and wanting to participate but not being able to were isolating and mentally difficult experiences. Overtime, the many challenges of dealing with hemophilia and its impact on their lives accumulated substantially, as a man in the 40 years and older group expressed it:

I realized in the past, you know, 5 or so years my hemophilia obviously effects me as I physically move through the world, but it has also had, a fairly profound effect on how I move through the world emotionally, and that I spent a great many years focusing on the gift that hemophilia was and the gift that HIV was. But that’s really only half the story, there’s another side, that living with the pain and the virus wore on me, so yeah, I think that [of] the psychosocial aspects that are at work.

Although self-infusion and prophylaxis were advancements in hemophilia management, from the psychosocial perspective they also made it difficult to adhere to treatment because essentially every treatment was another reminder of having the disorder, of being vulnerable: “every time you do [infuse, take a pill for HIV], you’re in that space of ‘I have these things’.”
As a man in the 40 years and older group who acquired HIV from factor use explained of getting tired of being someone with hemophilia and HIV, “so if you don’t take that pill, or you don’t use that needle, I think there’s something subconsciously going on that it’s like: ‘I’m not this’.”

Despite having been aware of missing treatment more often than he should, this was his way of pushing back against hemophilia which resulted in daily prophylaxis regimen becoming five to seven infusions a week instead. These experiences suggest that protest masculinity was present in men’s hemophilia management behaviours. Instead of using alcohol, drugs, violence, or crime, study participants delayed treatment, not infused prophylactically, or participated in high risk physical activities to demonstrate their masculinity, behaviours consistent with protest masculinity. Although men in the 40 years and older group were most articulate about the psychosocial aspects of the disorder (e.g., experiencing depression, being emotionally worn out) and pushing back against hemophilia, participants across all age groups described similar experiences.

Not showing vulnerability and weakness was another commonly used strategy by men to downplay or conceal their hemophilia. Although men found that most people were receptive to hemophilia, some stigma continued to surround it and all men had experienced stigma first hand. The stereotypical misconceptions portrayed men with hemophilia as very fragile. Several participants shared experiences of other people remarking: “if you get a paper cut will you die?” or “if you get punched will you die?”. These perceptions were contrary to masculine ideals of strength and toughness and therefore were perceived on the receiving end as undermining their male identity. Moreover, implicit was the suggestion of fragility round the men’s bodies wherein the protection from others – a scenario synonymous with female bodies – was evident. Hence, men had to decide when and how to disclose their disorder to others.
Many participants avoided disclosing their disease and only shared it with people that needed to know or that were close to them: “I’ve always had a bit of an attitude, maybe for better or worse, of not wanting anybody to pay attention to my hemophilia. I don’t like feeling different. And as a result I don’t share a lot about it.” Some men even averted bringing the attention of their loved ones to their bleeding episodes for the same reason. Disclosing hemophilia to strangers, some men were simply tired of giving long explanations others avoided the use of words like “disease, injection, shoot up” as these words had a negative connotation and hemophilia was misunderstood. Therefore, many of them disclosed their condition only strategically:

I explain to the workplace. I tell them it’s like a cold, I get a bleed every once in a while and then that’s it. I don’t go into details, I just give them the gist of how often I might need time off work. It’s easier to understand that way than explaining all the theories.

Many shared the knowledge that they had hemophilia with others gradually as the relationship evolved whether the relationships were friendships or intimate relationships. Often potential partners were well aware before the relationship progressed to that stage. However, a man in the 25 to 39 years old group found it beneficial to disclose hemophilia up front: “I found it’s a good measuring stick to see what that person is like.” The response to the disclosure helped him decide whether to further pursue the relationship or not. Only few men reported telling anyone about hemophilia: “I’m open. I’ll tell anybody,” or “I’ve always kind of told everybody.” It is interesting to note that only those who reported being willing to disclose hemophilia to anyone were using daily prophylaxis. This may suggest that there may be a connection between openness about hemophilia and men’s hemophilia management decisions.
However, findings suggest that there are no connections between men’s willingness to disclose their disorder and age sub-groups.

4.1.2.4 Establishing a Healthy Masculine Identity

Men presented an extraordinary resilience toward both physical and emotional limitations the disorder placed on them by continuously working toward overcoming those limitations. They placed a high importance on independence and autonomy in managing their disorder, an aptitude consistent with masculinity. Navigating the physical activities and treatment regimens in the context of constructing their masculine identity was a continuous process. This process often involved “pushing back” against hemophilia, a form of protest masculinity. For periods of time, some men were able to reach a harmonious state of being, a “healthy” balance between hemophilia limitations and their masculine identity, as a man in the 19 to 24 years old group explained: “some days you don’t even think that you have any disease at all because you can be so self-sufficient.”

Having a supportive environment where men were able to openly share about their disorder and their needs without being ridiculed or being made feel inferior was essential for finding that harmonious state of being. The participants identified three main lessons learned in their hemophilia management journey including: disclosure, taking care of one’s body, and don’t let it hold you back. Disclosure didn’t just mean telling others about hemophilia, but letting others know when one needed help. Taking care of one’s body meant navigating the physical activities and treatment options to minimize the number of bleeding episodes over a specific period of time. Don’t let it hold you back meant that “if you manage it [hemophilia] properly you can do anything you want to, really.” However, disclosure could also be perceived as showing one’s vulnerability which is contrary to masculine aptitude of stoicism. Looking after
one’s body or health is also not considered a masculine behaviour, on the contrary, men showing no concern for their health or wellbeing are considered strong and fearless. Therefore, men who did not feel comfortable with disclosure or looking after their body may have focused on doing what they wanted and risked injuries.

To find a healthy balance between masculine identity and hemophilia, men needed to muster alternatives to physical strength and other masculine attributes. Discipline, control and rationality in effectively self-managing their illness afforded important masculine capital in this regard: “It’s all based on logic, right? I’d look at the situation, assess the situation then make a choice.” The same man also reported developing his own software to track his treatment regime and strictly infusing every three days based on plasma levels and experiencing only three bleeding episodes a year. This man experienced the accomplishment of such control over his disorder as a major achievement that he was proud of.

Although generally men preferred infusing as rarely as possible, some were committed to daily prophylaxis to be able to have a physically active lifestyle, for example working in construction or playing sports. The concept of hierarchy of threats to one’s masculinity may explain why some men were more likely to seek help, adhere to strict treatment regime, adjust the level of physical activity, and look after one’s body which in return enabled them to get fulfillment through work, sports, family or other aspects of their lives. Those men who recognized the interdependence between their ability to achieve personal goals and hemophilia management practices were more likely to engage in self-preserving practices. It is interesting to note that those participants who presented a more balanced approach to managing their hemophilia also reported the least number of bleeding episodes in a year (ten or less). This may
suggest a connection between effectiveness of managing hemophilia and developing a healthy masculine identity.

4.2 Summary

Considering age differences in men’s experiences of hemophilia, findings suggest that men in the 40 years or older group were most limited physically due to the advancement of joint damage, most impacted by infectious diseases acquired through factor concentrates in the 1980s, most likely to perceive prophylaxis as a treatment with limited benefits, and became experts at managing their disorder through experience. In contrast, men in the 19 to 24 years old group were still learning to manage hemophilia, most likely to express the need for peer support opportunities, and most like to be physically active and engage in regular exercise. Older men perceived that recent advancements in hemophilia care (e.g., prophylaxis and regular exercise regimes) provided limited benefit for them because of the joint damage already done and would really benefit from addressing joint disabilities. While the younger generations benefited the most from recent advancements in hemophilia care, there is a need for further improvement in optimizing their health.

In addition, study participants were in a subordinate or marginalized masculine position because of the physical and psychosocial impacts of their disorder. Particularly around the age of high school years, a critical time of establishing a man’s masculine identity, study participants worked hard at establishing an identity not predominantly defined by their disorder. Overcoming their limitations and establishing their identity was a complex and delicate process that often included testing the limits of their bodies, disregarding the need for treatment, and engaging in unnecessary risky physical activities. Virtually all of them protested against their condition at some point in time. Results suggest that the development of albeit modified but balanced and
healthy masculine identity empowered them to surmount the limitations, minimize injuries, and likely improved health status and life quality over time.
Chapter 5: Discussion

Qualitative description and grounded theory methods were used to describe men’s experiences with severe hemophilia and inductively develop the findings according to age differences and in the context of masculinities. Findings suggest that age differences and masculinities play an important role in men’s experiences of severe hemophilia and may impact hemophilia management decisions.

5.1 Masculinities and Hemophilia Self-Management

One of the main findings from this study was that socially determined masculine ideals and behaviours play an important role in men’s identity throughout their lives, but have the strongest influence during adolescence, a critical time period in the development of individual identity, a process that can extend into young adulthood. Although other research has identified masculinity as an important factor in men’s hemophilia management decisions (Park, 2000), this is the first study to explore men’s experiences with hemophilia through a masculinities lens. Findings suggest that masculinities impact men’s experiences of severe hemophilia and even hemophilia management decisions. Men’s account of their experiences with severe hemophilia provided compelling evidence for typical men’s health related behaviours: stoicism, delaying treatment, denying or ignoring injury and potential consequences. Men reported taking unnecessary physical risks that resulted in injuries so that they could meet their psychosocial needs (e.g., not treating immediately a sports-related ankle injury) is consistent with masculinities literature suggesting men engage in self-destructive practices in the pursuit of embodying masculinity (Courtenay, 2000). In addition, severe hemophilia also impacted men’s identity and self-perception. When men sensed that severe hemophilia thwarts their ability to embody masculine ideals and behaviours, men may have pushed-back against hemophilia, a
form of protest masculinity and a behaviour that can impact their short and long-term health status. In other words, the more men felt limited by hemophilia (in physical activities, social or work opportunities), the more likely they were to rebel against hemophilia regardless of long-term consequences. Constantly facing limitations imposed on them by the disorder created a strong desire for a sense of “I am not this,” even if only for a short period of time. From a masculinities theory viewpoint these protests might be understood both as denying the marginalization invoked by hemophilia and aspiring to hegemony despite the disabilities that accompany this disorder. Therefore, it is critical to address not just the physical aspects of the disorder (treating or preventing bleeds), but also the variety of limitations the disorder can place on men.

Although for the most part men are typically reactive to health related issues (Courtenay, 2000), study participants emphasized independence, autonomy and self-reliance, which are important masculine attributes (Connell, 2005). Furthermore, participants specifically expressed having control over hemophilia and managing it on their own (with guidance from the clinic, but without influence by others) which is consistent with the masculinities concept of men conquering nature. Said another way, some masculine ideals can be mobilized to garner optimal hemophilia management practices. The implication of this finding for clinical practice is the importance of not trying to change men in the context of hemophilia, but to collaboratively work with men to enable them to overcome or at least minimize the limitations severe hemophilia imposes on them while affirming them in the masculine virtues that can catalyze effective self-management.

Clearly, some aspects of their masculine identity can be so important to them that it motivates them to commit to excellent hemophilia management practices. For example, men can
follow daily prophylaxis and/or regular exercise regimens in order to participate in sports activities, perform masculine jobs (i.e., construction), or develop a masculine physique (by working out at a gym), a phenomena consistent with the concept of *hierarchy of threats* in which men with prostate cancer would exchange sexual prowess for survival so that they can still be a family provider (O’Brien et al., 2005). Indeed, men can find a healthy balance between their masculine identity and hemophilia when they understand the interdependence between achieving their personal goals and looking after their bodies. Finding a healthy masculine identity in the context of hemophilia means managing hemophilia in a way that optimizes their bodies which in turn enables them to pursue and achieve their personal goals, even if somewhat modified masculine goals.

Men presented an extraordinary resilience against the limitations in their motivation to not let hemophilia define them. In their pursuit of constructing an identity that is not defined solely by hemophilia, in the model of “I am not this”, by pushing back against hemophilia men tended to engage in hemophilia management practices that had negative short and long-term consequences. On the other hand, if men perceived hemophilia management practices as a means to achieve their personal goals, men were motivated to engage in optimal hemophilia management practices and established a healthy masculine identity.

In this study participants indicated that pain, and not earlier symptoms of bleed, is the cardinal sign that prompts men to take action. This is consistent with wider men’s health literature; for example, in cardiac health White and Johnson (2000) suggests that in order for men to fulfill society’s expectation of being strong and productive, men need to experience substantial discomfort and debilitation before they seek help. In the current study, participants reported delaying treatment until the end of a sports activity or going to sleep at night without
treatment despite being aware of a potential bleed. This is consistent with men’s denial and rationalization of symptoms as seen in other men’s health research (White & Johnson, 2000) for fear of being seen weak or facing the possible consequences of one’s vulnerability. Both rationalization and denial result in delaying treatment. Older participants were more likely to state that after experiencing substantial bleeding episodes, they learn to recognize bleeding episodes, activities likely to result in a bleeding episode, and effective treatment strategies. Even though men would become proactive in managing hemophilia, they remain reactive and not proactive towards other health concerns.

Although there are earlier signs of bleeding prior to pain and joint swelling, such as tingling and warmth in the joint, participants didn’t refer to relying on those signs to initiate treatment. This is an important finding to consider when planning for better enabling men with severe hemophilia to treat bleeding episodes and minimize joint damage. However, it is not clear from participants’ accounts whether or not they are aware of other earlier signs or simply rely on pain only. This needs to be further studied and clinical staff should consider assessing each man’s individual knowledge of early signs of bleeding episodes.

5.2 Transition to Adulthood

Participants across all age groups reported experiencing high incidence of bleeding episodes that resulted in joint damage during their high school years. Although participants in the 19 to 24 years old group verbalized knowing their body and controlling hemophilia well, they also sought out opportunities to share and confirm their experiences with others. In addition, with the exception of one man on daily prophylaxis, study participants in the 19 to 24 years old sub-group reported the most bleeding episodes in a year (from 20/year to as high as 4-6/month) among all study participants. These findings suggest that experiencing high incidence of
bleeding episodes during adolescence is extended into young adulthood and is consistent with other hemophilia literature that reported highest bleeding incidences among patients 13 to 24 years of age (Zappa et al., 2012). In other words, adolescent and young adult males are still working through the process of mastering hemophilia management and more support is required to enable them to reach mastery sooner. Preventing bleeding episodes during this time period is critical for improving long-term health outcomes as even a few bleeding episodes can result in irreversible joint damage (Manco-Johnson et al., 2007).

These challenges are not unique to hemophilia, as literature suggests similar behaviours with other chronic illnesses, such as diabetes and cystic fibrosis (Di Battista, Hart, Greco, & Gloizer, 2009; Towns & Bell, 2011). Moreover, recent literature suggests grouping together patients with chronic illnesses in the 10 to 25 years of age for health care related needs, instead of the traditional 12 to 19 years of age generally referred to as adolescence in pediatric health care settings (English, Park, Shafer, Kreipe, & D'Angelo, 2009). This is due to neurological development extending into the mid-twenties (Giedd, 2009). Furthermore, it is suggested that the preparation for the transition should begin a few years prior to transfer to the adult care program, ideally at 12 to 16 years of age (Towns & Bell, 2011). Disrupting these definitions Kimmel (2008) argues that most men endure a prolonged period of adolescence wherein they are at school longer and don’t marry and/or have a family until their late 20s. This shift has revealed 30 as the new 20, and in the context of dealing with young men who have hemophilia the expectations of age specific groups may also be changing.

Although literature suggests that adolescence is a critical period in hemophilia care and health care providers have made efforts to improve care during this period, further improvement in the transition of care is suggested by the literature (Breakey, Blanchette, & Bolton-Maggs,
2010). Further research and innovation in the critical time of adolescence and young adulthood should include a developmental perspective (from childhood through adolescence to adulthood) and transferability of lessons learned in other chronic illnesses should be assessed (Krasnegor, 1993). Findings of this study suggest that in addition to a developmental perspective, masculinities should also be included to further improve hemophilia transition of care.

5.3 Treatment and Self-Management

Although previous research reported that the use of prophylaxis decreases with age (Zappa et al., 2012), it offered no explanation for that phenomenon. The suggestion by older study participants with advanced joint damage in this study that no amount of prophylaxis will correct their disability and significantly improve their mobility may partially explain that phenomenon. Older participants shared their belief that prophylaxis is too late for them as they already have significant disability. Their advanced joint damage limits their physical activity to walking and no amount of prophylaxis will reverse that.

In addition, the finding that the participants perceived that the relatively short half-life of factor (less than 24 hours) limits its practicality for prophylactic use is a new and may be a significant finding in understanding the lag in wider prophylaxis uptake. Indeed, men in all age stratified sub-groups expressed the desire to reduce the frequency of infusions with prophylaxis. The concern over long term venous access, disliking performing frequent self-infusions, and that frequent infusions (i.e., daily prophylaxis) serves as a reminder of their disorder creating a psychological barrier to prophylaxis is consistent with and confirms the findings of previous hemophilia research (Lindvall et al., 2012; Remor, 2011) that suggested these findings act as barriers to wider use of prophylaxis.
Most men understood that prophylactic treatment is the most effective method to prevent bleeding episodes and joint damage in the long term as suggested by previous research (Manco-Johnson et al., 2007). The finding that experiencing more bleeding episodes and advancement of joint damage motivates men to consider prophylaxis to slow exacerbation of symptoms is also consistent with other research in hemophilia (Llewellyn et al., 2003) which suggested that men’s perception of necessity of treatment improves adherence to treatment. The findings of this study both confirm the results of previous research and add additional concerns surrounding prophylaxis. Findings suggest that men need help in overcoming barriers in order to achieve a wider uptake of prophylactic treatment.

Men expressed that regular exercise can help with preventing joint injury, a finding consistent with previous research and recommendations, which have suggested that regular exercise can strengthen joints and minimize bleeding episodes (Czepa et al., 2013; Lane et al., 2013; Negrier et al., 2013). However, some participants reported lacking the knowledge to conduct strengthening exercises safely that result in improved physical condition without causing bleeds. This finding is consistent with previous literature (Negrier et al., 2013), which also suggested that there is a need to continue to provide education and support on how to conduct muscle strengthening exercises safely and regularly. Lastly, the finding that participants’ understanding of the benefits and purpose of joint braces (i.e., ankle braces) is different than the intended clinical purpose is intriguing. Overall, these findings suggest that comprehensive education and support is needed for improving men’s physical fitness in a safe and effective way resulting in long-term health improvement.
5.4 Social Engagement and Support

The physical limitations severe hemophilia imposed on men were significant, but the impact on other aspects of their lives such as social engagement, travel and work or career options were not negligible either. This is consistent with previous literature which suggested that in addition to providing education on causes and consequences of bleeds, bleed prevention and treatment, counseling needs to be provided on the psychosocial aspects of the disorder, such as career selection, travel and disclosure (Lane et al., 2013). The development of heightened vigilance for physical danger and the reality of physical limitations due to severe hemophilia can result in the development of an inferiority complex and self-limiting mentality, a finding consistent with previous research (Negrier et al., 2013) which suggested that men with hemophilia may be reluctant to engage in regular exercise because of it. Men with severe hemophilia may not be able to compete with others in some sport or physical activity, enabling them to find other physical activities where their masculinity can be expressed safely would facilitate the development of a healthy masculine identity. Although previous literature suggested educating men on risks of different physical activities (Lane et al., 2013; Negrier et al., 2013), these findings suggest that for the most part men are able to assess the risks associated with specific physical activities but decisions to participate are made in the context of individual and social needs and preferences. Men need education on effective strategies to minimize the risk of injury while participating in riskier activities.

Their social engagement was also influenced by hemophilia as men with severe and moderately severe hemophilia were likely to engage in physically less active but supportive social settings without having to experience undue bias or presumptions. Many of them were cautious about when and how to disclose hemophilia, providing education, counseling and
positive experiences is needed to enable men to overcome this difficulty. Opportunities to engage with peers who have hemophilia were valued and sought after (i.e., camps, meetings, outings), as suggested by previous literature (Breakey et al., 2010; Lane et al., 2013) which suggested that peers can provide encouragement and act as role models. However, men also expressed frustration with the limited means to connect with other men with hemophilia outside of these events. The effectiveness of current opportunities for peer support should be re-assessed and additional opportunities considered with the needs of men with hemophilia in different age sub-groups in mind. These opportunities should include same age peer engagement, but mentorship and coaching partnerships may also be valuable for both parties and be an excellent strategy for knowledge transfer. Lastly, their work and career options were also limited by severe hemophilia with both short and long term consequences. Further opportunities to maximize individual employable skills need to be developed which may include counseling on appropriate career choices at a young age or arranging financial assistance for skills re-training for older men.

5.5 Limitations

Replicating the study in different populations is needed to confirm the findings. Further research is required to better understand the connections between masculinities and men’s experiences of severe hemophilia.

5.6 Recommendations

It is apparent that the availability of safe factor products, the practice of self-infusion and prophylaxis along with other care improvements over the last couple of decades have translated into improved physical mobility and overall health for men with severe hemophilia. However, results suggest that there is still plenty of room for improvement in hemophilia care to minimize
limitations and improve overall health. In addition to treating or preventing bleeding episodes, there is a need for a comprehensive treatment approach that enables men to effectively manage hemophilia in the complex context of their lives. Along with physicians, hemophilia care nurses always played and will likely continue to play a central role in care management because of their close connections with their patients. Integrating other health care providers into the care conundrum, such as social workers and physiotherapists, is vital to establish a comprehensive care approach.

5.6.1 Recommendations for Nursing and Clinical Practice

Results support the recommendations by previous research for providing individualized education (Lane et al., 2013) throughout men’s lives to optimize hemophilia care including but not limited to recognition of bleeds, effective treatment and prevention, consequences of bleeds and lack of treatment, assessment for and reduction of risks for joint injuries, planning for travel and work, among others. However, findings that suggest men are unfamiliar with the early signs of joint bleeds and perceive the purpose of joint braces as joint protection instead of gait support highlight that men’s knowledge should not be assumed and need to be continuously assessed.

Nursing care should include the assessment and teaching of even the basics in hemophilia care, such as early recognition of bleeding episodes that is based on symptoms other than pain, swelling or loss of range of motion. As suggested by others, standardized learning needs assessment tools may aid in this (Lane et al., 2013).

Findings suggest that masculinities are a powerful context in which men experience severe hemophilia and have to navigate hemophilia management practices. The limitations imposed on men’s lives by hemophilia need to be assessed and educational and treatment needs individualized in that context. Although men may not be the experts in some clinical aspects of
hemophilia, they are the experts on how hemophilia impacts their lives. Helping men to overcome those limitations will more likely engage them and commit and focus their efforts to optimize their lives through optimizing their health. In terms of clinical practice, nursing as a predominately female occupation should be attuned to their gendered role in co-constructing masculinities with their male hemophilia patients. Specifically, by affirming men, especially young men, to honestly share their management strategies in relation to their activities, nurses may be able to positively influence men’s practices. Inversely, the findings from the current study suggest that behaviorist approaches to patient education and hemophilia management are more likely to yield hyper masculine ‘risky’ performances than treatment compliance. Related to this, nurses have unique opportunities to influence men toward strength-based self-management strategies by working with rather than to necessarily change men’s health practices.

Findings imply that the emotional and psychological aspects of hemophilia has profound effect on men and that some experience depression but are hesitant to talk about it, a behaviour consistent with literature on men’s mental health (Oliffe & Phillips, 2008). Incorporating counseling and social work services into hemophilia care should enable men to cope with the emotional and psychological aspects of the disorder, but integrating stress reduction strategies and assessment for depression should be considered.

In addition, as much as education on the benefits of regular exercise and reducing risks of participating in physical activities is important, some men may require more to effectively incorporate that knowledge into their lives. For example, older men may require the direct involvement of personal trainers in a safe environment initially to consider regular exercise in the first place. Younger men may be more knowledgeable about regular exercise but are also more likely to push their physical limits to embody masculinity. Providing opportunities for sports
activities that men with severe hemophilia can participate in safely, challenge themselves physically and demonstrate the competitive aspect of masculinity may further reduce risks for injuries. This doesn’t necessarily have to be limited to the hemophilia community but may include the general population.

Developing a well organized peer network that creates a community feel for men can create a supportive and safe environment where experiences and knowledge can be shared and enable men to cope easier with challenges. Although not all men at all times would require it, a supportive community can be particularly important at a younger age. In addition to formal events organized by CHS or care providers, there is a need for an informal network where men can connect with others more readily. Lastly, collaboration between the pediatric and adult programs is essential not only to ensure a smooth transition of care but from a developmental perspective from adolescence into adulthood.

5.6.2 Recommendations for Nursing and Healthcare Policy

The definitions of prophylaxis (i.e., continuous vs intermittent, primary vs secondary) are inconsistent both in the literature and in men’s understanding of it. This makes it difficult for men to comprehend the potential benefit of a particular prophylaxis, yet research suggests different benefits for continuous and intermittent prophylaxis (Manco-Johnson et al., 2007). Applying standardized prophylaxis definitions in clinical practices would make the comparison of research results easier and may help with men’s understanding of the actual benefits.

The study findings suggest the need to continue to fund and improve comprehensive hemophilia care based on a multidisciplinary approach. In addition, the benefits of peer networking opportunities and developing a hemophilia community involvement in planning hemophilia care and funding should be prioritized.
5.6.3  **Recommendations for Research**

Considering that this is the first study that explores hemophilia from the masculinities perspective, further research is required to build upon the findings and evaluate the transferability to other hemophilia populations. Confirmatory analysis with participants who are not from a West-coast or urban environment would be beneficial to verify the findings as men in other parts of the country or the world and rural settings may have different experiences with hemophilia. Although findings suggest connections between masculinities and men’s hemophilia management practices, further research is required to better understand these connections.

Findings highlight that adolescence and young adulthood is a critical period in hemophilia management practices, a finding consistent with literature in other chronic illnesses during this period. Findings also suggest that masculinities play an important role during this period. However, further research is required on how to help adolescents with hemophilia transition into adulthood while optimizing health status. Studying hemophilia in adolescence and young adulthood from the developmental theory perspective may provide additional knowledge on this critical time period.

Results indicate the need to develop factor products with longer half life because such factor products would likely improve use of prophylaxis and overall health outcomes. However, men’s reluctance to be exposed to new products need to be effectively navigated to ensure successful uptake of new products. In addition to improving prophylactic treatment, findings suggest the need to further research the benefits of other strategies to prevent joint injuries, such as regular exercise and use of joint braces. Particularly for older men with advanced joint damage, further research on possible methods of correcting joint damage (i.e., joint replacement,
chondrogenesis with stem cells) would be beneficial to identify the most effective methods. Although ankle fusion is one option for decreasing pain from severe ankle damage, the options for correcting other damaged joints is limited.

5.7 Summary

Findings suggest the need to further improve comprehensive hemophilia care that is individualized based on one’s trajectory with this chronic disorder. There are some significant overall differences in needs based on age. For the youngest generation, transition of care is a critical period to prevent future health problems, which may range from as early as age 10 to as late as age 25 years old. Education on physical aspects of the disorder, bleed prevention, treatment and benefits of regular exercise (among others) and counseling on psychosocial aspects of the disorder is most warranted. Providing opportunities for peer support can also be vital to better support men with severe hemophilia. Older men would likely benefit the most from improvements in addressing advanced joint damage, chronic pain, and disability. In addition, their perceptions that prophylaxis is not going to benefit them should be further explored. Furthermore, improving and sustaining their mobility through regular exercise should improve their quality of life and prevent other age related health issues.

Physical limitations caused by severe hemophilia can impact most other parts of men’s lives including emotional and social aspects. Findings suggest that socially constructed masculinities serve as an important context in which men experience and manage severe hemophilia. The connections between masculinities and severe hemophilia impact the development and construction of one’s masculine identity. The construction of masculine identity can hinder hemophilia management practices. However, men are more likely to optimize hemophilia management if able to develop a masculine identity nourished by
interdependence amid optimizing their health and achieving their personal goals. This research provides a new perspective on men’s experiences with severe hemophilia. Although further research is required to confirm the validity of this perspective, it should be considered to continue to improve hemophilia care.
References


Oliffe, J. L., Kelly, M. T., Bottorff, J. L., Johnson, J. L., & Wong, S. T. (2011). "He’s more typically female because he’s not afraid to cry": Connecting heterosexual gender relations and men’s depression. Social science & medicine, 73(5), 775-782. doi: 10.1016/j.socscimed.2011.06.034


### Appendices

#### Appendix A  Demographics

<table>
<thead>
<tr>
<th>Employment status:</th>
<th>Marital status:</th>
<th>Education level</th>
<th>Where do you live?</th>
</tr>
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<tr>
<td>□ Student</td>
<td>□ Single</td>
<td>□ High School</td>
<td>□ Urban (e.g. BC Lower Mainland)</td>
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<td>□ Working</td>
<td>□ Married</td>
<td>□ Diploma or Certificate</td>
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<td>□ Live w/partner</td>
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<td>□ Retired</td>
<td>□ Divorced</td>
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<tr>
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<td>□ Widowed</td>
<td>□ Master’s degree</td>
<td></td>
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<td></td>
<td>□ Other, specify</td>
<td>□ Doctoral degree</td>
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</tr>
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<td></td>
<td></td>
<td>□ Other, specify</td>
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<tr>
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<td></td>
<td></td>
<td>□ Rural</td>
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<table>
<thead>
<tr>
<th>Type of hemophilia:</th>
<th>□ A (Factor VIII deficiency)</th>
<th>□ B (Factor IX deficiency)</th>
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</table>

<table>
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<tr>
<th>Inhibitor history:</th>
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<th>□ Yes, in the past</th>
<th>□ No</th>
<th>□ Unknown</th>
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</thead>
</table>

<table>
<thead>
<tr>
<th>Type of current treatment</th>
<th>□ on-demand</th>
<th>□ prophylaxis</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>Have you started prophylaxis for the first time within the last 3 years?</th>
<th>□ Yes</th>
<th>□ No</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>Have you been seen in a hemophilia clinic within the last 1 year?</th>
<th>□ Yes</th>
<th>□ No</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>Estimate how many spontaneous (non-traumatic) bleeding episodes you have:</th>
<th>_____/month</th>
<th>_____/year</th>
</tr>
</thead>
</table>
Please circle the joints in which you have damage from hemophilia.
Appendix B  Focus Group Guiding Questions

B.1 Initial Guiding Questions

1. When do you use factor?
   a. Probe – how would you describe your overall approach
   b. Probe – re-use participant language to clarify and detail how the men differentiate and operationalize prophylaxis (i.e., long-term, intermittent use) and on-demand factor

2. How do you weigh up risk (vs. benefit) when deciding if and how to use factor prophylaxis?
   a. Probe – if you use prophylaxis, when do you break your own ‘rules’ about using factor preventatively?
   b. Probe – what is your major concern about using factor preventatively?

3. How has, and how do you expect your use of factor to change as you age with respect to prophylaxis?
   a. Probe – What have been/do you expect to be the biggest challenges?

4. Tell us about the people who most influence your decisions around managing your hemophilia.
   a. Probe – delineate if non-hemophilia or hemophilia peers, family members, health care providers, etc. – and clarify context by soliciting examples about the most influential people in men’s Hemophilia management.
   b. In terms of health care providers (doctors and nurses) who most influences your decisions?
      i. Clarify gender of provider in asking how that relationship works – or not
      ii. Women often look after men’s health – how does that play out for you?
      iii. Follow-up – what is unique about your situation?

5. What activities most influence your management of Hemophilia (and decisions about using prophylaxis)
   a. Probe – build on answer and introduce other activities – i.e., what about – work, social activities, travel, leisure, sports – prompt to have participants expand on specific examples

6. Where do you access information about hemophilia? How do you evaluate that information? Who do you connect with online around your Hemophilia? Who would you like to connect with online around your Hemophilia?
   b. Probe – in person support groups, clinicians, friends, family?

7. Tell us how hemophilia influences your roles and relationships?
   a. Prompts around specific males roles and relations – father/uncle/brother/son, work intimate relationships, friendships, social activities

8. What are your three most important pieces of advice for other men who have Hemophilia?
   a. Probe – what resources would enable men with hemophilia to do better

9. What questions should we ask other men who have Hemophilia?
10. Any final thoughts – additional things we should tell healthcare providers about men who have Hemophilia?

B.2 Revised Guiding Questions

The Initial Guiding Questions were revised after the first focus group session to facilitate the flow of the discussion and to address additional relevant aspects of their experiences and perspectives on hemophilia.

1. What were the major milestones in your understanding and/or management of your condition?
   a. Probe – type of Hemophilia (severe, A or B)
   b. Probe – when was treatment started, how treatments changed over time, history with HIV/Hep C, changes in physical activity

2. How would you describe your use of the factor?
   a. Probe – how would you describe your overall approach
   b. Probe – re-use participant language to clarify and detail how the men differentiate and operationalize prophylaxis (i.e., long-term, intermittent use) and on-demand factor

3. How do you decide on when and how to use the factor? Please give examples.
   a. Probe – if you use prophylaxis, when do you break your own ‘rules’ about using factor preventatively?
   b. Probe – what is your major concern about using factor preventatively?

4. How has, and how do you expect your use of factor to change as you age with respect to prophylaxis?
   a. Probe – What have been/do you expect to be the biggest challenges?

5. How do different activities influence your management of Hemophilia (and decisions about using prophylaxis)
   a. Probe – build on answer and introduce other activities – i.e., what about – work, social activities, travel, leisure, sports – prompt to have participants expand on specific examples

6. Where do you access information about hemophilia? How do you evaluate that information? Who do you connect with online around your Hemophilia? Who would you like to connect with online around your Hemophilia?
   b. Probe – in person support groups, clinicians, friends, family, uncle, cousin?

7. Tell us how hemophilia influences your roles and relationships?
   a. Prompts around specific male roles and relations – father/uncle/brother/son, work intimate relationships, friendships, social activities, stigma, career, education.

8. Who most influence your decisions around managing your hemophilia?
   a. Probe – delineate if non-hemophilia or hemophilia peers, family members, health care providers, etc. – and clarify context by soliciting examples about the most influential people in men’s Hemophilia management (mother, nurses, brother, wife).
   b. In terms of health care providers (doctors and nurses) who most influences your decisions?
i. Clarify gender of provider in asking how that relationship works – or not
ii. Women often look after men’s health – how does that play out for you?
iii. Follow-up – what is unique about your situation?

9. What are your three most important pieces of advice for other men who have Hemophilia?
   a. Probe – what resources would enable men with hemophilia to do better
   b. Probe - management strategies, on-demand vs prophylaxis, exercise, prevention of injuries, impact on health and life as a whole

10. What questions should we ask other men who have Hemophilia?
11. How does your “health related wisdom” gained through hemophilia influence the way you manage other health issues?

12. What would your suggestions be to parents of kids with hemophilia?
13. What could the hemophilia clinic staff do to improve the care provided?
   a. Probe – what works well, what could be improved
14. Any final thoughts – additional things we should tell healthcare providers about men who have Hemophilia?

B.3 Sub-Appendix

This is Appendix B, Section 1.