Hemophilia Treatment Accessibility: An Analysis of Unexplored Barriers

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Abstract

In the research conducted, the accessibility of treatment for the bleeding disorder hemophilia is explored. While current care for people with this genetic anomaly is available, there are additional concerns and variables surrounding the medical advancement and treatment which should be considered not only by health professionals, but also in the medical laboratory setting. This thesis asks what the problems and hindrances with the generalized path of care prescribed are, and with the knowledge acquired, it pursues to recognize the under-discussed barriers to hemophilia and how to better advance the future care of hemophilia patients by creating a basis of discussion to these barriers for future and current healthcare professionals' use. With this uncovered information, it is expected that the understanding of the potential hardships with hemophilia care is furthered, and while the analysis strives to ultimately benefit people with this disorder, it also seeks to encourage healthcare professionals to gain an improved understanding that the knowledge of diversely accessible treatment options is essential in the workplace.

keywords: hemophilia, HTCs, blood disorder, adolescents, women with hemophilia

Introduction: *What is Hemophilia?*

Hemophilia is a X-linked recessive genetic disorder that impacts a process called hemostasis, which is the property of blood circulation that keeps blood as a fluid within blood vessels under normal circumstances and prevents blood loss due to injury (McKenzie, 2009). People with this condition either completely or partially lack the essential blood-clotting factor VIII (hemophilia A) or factor IX (hemophilia B) due to mutations on either the F8 or F9 gene on the X-chromosome, and these factors are proteins in the blood that take part in what is known as the coagulation cascade. The components of the coagulation cascade come together to create a blood clot by the formation of the desired end-product, cross-linked fibrin. In addition to being an inherited disorder, hemophilia can also be acquired through either the creation of neutralizing FVIII antibodies from the use of transfused FVIII, or idiopathically. Though hemophilia A and B present identically, certain points in this analysis and the literature discussed may gravitate towards the aspects surrounding hemophilia A specifically since “Hemophilia A is about four times as common as hemophilia B” (CDC, 2020).

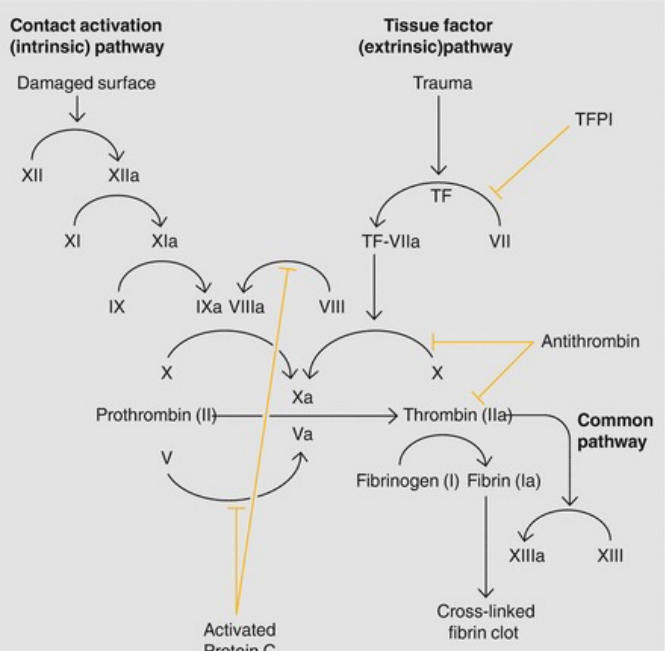
Hemophilia places people at high risk of “musculoskeletal bleeding involving the muscles and soft tissue”, joint bleeding, as well as prolonged bleeding following injury or surgery (Smith, 2019). These are hallmark signs of bleeding disorders of secondary hemostasis. Additional signs and symptoms are more dependent on severity of the disease. Normal functional activity of coagulation factors is 50 to 150% (or 50.0 to 150.0 IU/dL), so symptoms of hemophilia typically begin to show at 30% factor activity or below. Mild hemophilia is classified as FVIII activity between 30% and 6%, and spontaneous bleeding is usually not common in these cases. Moderate hemophilia is FVIII activity between 5% and 1% accompanied with bleeding due to trauma, dental work, injury, and surgery. Joint and muscle bleeding can be

commonly seen. Finally, any FVIII activity that is less than 1% is classified as severe hemophilia, which usually presents in the first few months of life as spontaneous bleeding, and resulting internal bleeding has the capability to impact internal organ functions.

Because hemophilia can cause bleeding in the joints, commonly in the knees, elbows, ankles, shoulders, and wrists, these areas become painful and swollen and there is an increase of incidence with age. Intracranial hemorrhage during the neonatal period is a severe complication that is a leading cause of death in hemophilia newborns, and potential brain bleeding in older patients leads to falling, confusion, and lethargy. Additional symptoms include blood in the urine (hematuria), bleeding into deep muscle tissues, blood leaking from a vessel and collecting below the skin (hematoma), and delayed bleeding due to the ability of primary hemostasis to create a platelet plug but the inability of secondary hemostasis to create the fibrin blood clot. Bleeding after circumcision and unexplained, easy bruising are also familiar symptoms of both hemophilia A and B. Due to inherited hemophilia being X-linked, more males tend to be symptomatic than females, and the number of reported men with hemophilia in a study done from 2011 to 2020 greatly outnumbers the number of females: 23,728 males and 3,504 females (Miller et al., 2020).

The process of coagulation is split into two pathways based on the biological properties of the clotting factors involved, as demonstrated in Figure A, and FVIII and FIX fall into what is

Figure A



(Shantsila & Lip, 2016)

known as the intrinsic pathway, which is activated by damage to the endothelial surface. Due to hemophilia being a problem with creating a fibrin clot, routine testing for hemophilia includes clot detection analysis. A measurement of the time it takes for a patient's plasma – where the coagulation factors reside – to form a fibrin clot is done by either automatic or semi-automatic analyzers that can be either optical or mechanical in sensory detection. These two factors' ability to clot in this pathway is evaluated by a test called the activated partial thromboplastin time (aPTT), which is based on the principle that in citrated plasma, the addition of a platelet substitute, factor XII activator, and calcium chloride allows for formation of a stable clot; then the time required for the formation of a stable clot is recorded in seconds (Ignjatovic, 2013). Low factor levels – within the intrinsic pathway – that impact the blood's ability to create a clot will prolong the aPTT due to the disruption of the coagulation cascade, therefore hinting there is a factor deficiency. Further evaluation utilizing specific factor assays can determine the specific factor deficiency, and a Bethesda assay, which is applied for the detection of FVIII inhibitors – neutralizing antibodies created from recombinant FVIII treatment – are also common instruments of evaluation.

An evaluation of the incidence and prevalence of hemophilia among US males using the national hemophilia treatment center network was conducted from 2012 to 2018, and results concluded that "we estimate that there are between 29,761 and 32,985 males with hemophilia living in the United States [as of 2020]" (Soucie et al., 2020).

Though the prognosis for many people with hemophilia is not as dire as it may seem to the uninformed, and the options for care are hopeful and have been proven to work successfully, people with this condition are still susceptible to the challenges relating to the ease of care for their health. Current treatment options for hemophilia are simply treatments and fail to also

provide solutions to any barriers to obtaining such care, whether it be mental, physical, financial, or time-related.

Hemophilia Treatment Centers

The main places for hemophilia care are hemophilia treatment centers (HTCs), and HTC multidisciplinary teams comprised of a hematologist, nurse or nurse practitioner, social worker and physical therapist who provide expert diagnostic, treatment, prevention, education, counseling, rehabilitation and care coordination services to improve the health of people with genetic bleeding disorders (Riske et al., 2020; Baker et al., 2012). In 2014, a nationwide US patient satisfaction survey of HTC services and care processes was conducted, and 62% of all 5006 responses came from people with hemophilia. Of these respondents, 96.3% reported being satisfied with HTC team members, services, and care processes (Riske et al., 2020). However, the majority of the participants in this review had only mild hemophilia, and this may underestimate the satisfaction with HTC of those with moderate to severe hemophilia, which presents more severely and be more challenging to monitor due to the spontaneity of its symptoms.

Additionally, barriers to accessing these centers remain largely at hand. One of these barriers not always considered in routine hemophilia care is financial barriers. Patients reported that the primary barrier to their seeking care in an HTC was “insurance does not pay for comprehensive care at HTCs” (Saxena, 2013). In a cross-sectional study of non-attendance among patients at a US hemophilia treatment center from 2010 to 2014, “patients with public insurance – compared to those with private insurance – may have inconsistent coverage, difficulties with renewing coverage, job insecurity and may be disproportionately burdened by copays and other medical costs incurred by clinic visits” (Barry et al., 2018). Though the results

of this study was conducted at only one HTC, it is feasible to conclude that the characteristic of consistent non-attenders at many other HTCs are those found to have public health insurance.

Distance to an HTC also remains a difficult obstacle to overcome, particularly for patients who live in rural areas, those with no means of transportation, and those with crippling physical disabilities. A research study from Zhou et al. (2011) showed that "three of the most frequently cited barriers for patients in seeking treatment at an HTC were 'distance to the center' (30.0%–43.8%), 'clinic hours were not convenient' (26.7%–37.5%), and 'transportation to center' (20.0%–31.3%)". Additional support for the conclusion that hemophilia treatment centers, while beneficial, are not always convenient for patients with hemophilia can be found in a 2006 analysis of the average distance of HTC from those patients' homes. Using the CDC-sponsored Universal Data Collection (UDC) surveillance program that includes demographic and clinical data on about 90% of patients using these HTCs for their care, and the knowledge of where over 130 nationwide HTC were located in the United States, it was found that 82% of the 12,397 patients living with hemophilia at the time lived an average of 68 miles from the nearest HTC. Needing to make several trips a week for transfusion of FVIII becomes inconvenient to those who not only live a considerable distance away from a HTC. A solution to this is the ability to practice administering recombinant FVIII at home.

The Emergence of Non-Factor Therapy for Hemophilia

The most widely-used and common treatment for hemophilia is the use of recombinant FVIII (rFVIII or rVIII), which is routine prophylaxis with FVIII to compensate for the lack of FVIII in the blood. The benefits of home infusion of anti‑hemophilic factor (FVIII) for hemophilia include improved adherence to prophylaxis, prompt treatment of bleeds, decreased time lost from work or school, fewer trips to the emergency room and hospitalizations, and

increased patient autonomy. Additionally, not only does home treatment allow patients to treat a bleeding episode rapidly, but it is also associated with several short- and long-term benefits, such as improved work and school productivity as well as reduced costs related to bleedings (Kavakli et al., 2010).

However, due to the half-life of standard half-life (SHL) FVIII, patients with hemophilia must inject themselves frequently (on average three times per week) to maintain plasma FVIII levels ≥ 1%. This represents a "substantial burden and offers limited options to tailor prophylaxis to an individual patient's needs, which may impact adherence" (Hermans et. al, 2021). Furthermore, "the self-management of bleeds via home infusion of clotting agent requires significant commitment by patients…and issues include finding a vein, using a nondominant hand, and dealing with fear of needles" (Teitel et al., 2004). When it comes to parental involvement for at-home care for hemophilia in their children, parents express their concerns with injecting intravenous due to a variety of reasons such as anxiety about potentially contaminated blood products, fearfulness of doing something wrong or causing their child pain, and being squeamish about delivery of injections (Oyesiku, 2010).

Though bypassing agents such as recombinant activated FVIIa (rFVIIa) and activated prothrombin complex concentrates (aPCCs) are useful in the treatment of hemophilia with inhibitors due to to aPCC containing activated coagulation factors and rFVIIa activating the extrinsic pathway (and therefore bypassing the need for FVIII or FIX), "they are not ideal for long-term prophylaxis due to their low efficacy, high cost, inconvenience and increased risk of morbidity", and "no laboratory assay is currently validated either to monitor the efficacy of bypassing agents, or to determine their optimal dose" (Rocino, 2017; Shapiro et al., 2018). One new, promising treatment for patients with hemophilia is through the use of non-factor therapies

such as emiciztumab. Emicizumab is a humanized, bispecific monoclonal antibody, restores the function of missing activated factor VIII (FVIII) by bridging FIXa and FX to facilitate effective hemostasis in patients with hemophilia A; emicizumab prophylaxis significantly reduced the rate of bleeding events compared with no prophylaxis in adults and adolescents aged over 12 years with hemophilia A with *and* without inhibitors in phase III clinical trials (Blair, 2019). It is the first non-factor replacement product to be approved for the prevention of bleeding episodes in patients with hemophilia A, and due to its extended half-life properties, it can be administered potentially as infrequently as once every 4 weeks (Blair, 2019).

Though there are promising outlooks for the use of emiciztumab, there are barriers that literature does not always emphasize. One is the ability to equivaleate emicizumab to a corresponding amount of FVIII. Due to emicizumab's dependency on FIX, it is thus not possible in an in vitro biochemical assay to establish a true FVIII equivalence for emicizumab because it will depend on how much FIXa is used in the assay, and though emicizumab mimics FVIII cofactor function to some extent, the classic 1-stage APTT-based assay is not useful to measure emicizumab levels because the APTT is normalized already at *sub*therapeutic emicizumab concentrations (Lenting, 2020). Additionally, there have been reports of thrombotic microangiopathies due to the use of emicizumab with co-administration of aPCCs, and there is the need for immune tolerance induction regimens to be analyzed using aPTT tests, which, as previously stated, cannot be analyzed when emicizumab is in effect (Aleman, 2018).

Though the barrier surrounding the use of emicizumab is not with the validity of the non-factor therapy itself, it is understandable how the restriction of its use due to the inability to completely accurately administer and monitor amounts can be disappointing to those with hemophilia, especially since emicizumab is such a promising treatment. There is the hope that

specific assays for emicizumab can be created to "better connect ex vivo procoagulant activity with in vivo efficacy" (Aleman, 2018). These developments represent a paradigm shift in how we approach treating and monitoring patients on non-FVIII treatments, and the willingness of to put forth the required heightened degree of awareness and training for patients, clinicians, and researchers alike will help further the wellness for people with hemophilia and assist in their ability to lead lives with less risk for bleeding episodes.

Hemophilia in Young Adults

Like any other disease, patients with hemophilia undergo not only physical burdens but also carry emotional weight. Educational information and literature about hemophilia to maintain the condition, while vital for the health of those with hemophilia, does not always take into account the emotional burden that comes with living with hemophilia. Pinto et al. (2018) recognizes that though there is a considerable amount of literature examining illness perceptions, "studies in hemophilia field have only focused on their association with clotting factor treatment adherence therapy" rather than the mental health of people with hemophilia. Treatment of hemophilia can lessen the physical difficulties, but the reality is that inherited hemophilia is a life-long responsibility, which can manifest as a mental barrier to care for hemophilia by itself. Because of the rarity of the X-linked disease, it can be challenging for youth to create bonds with others who can fully relate to their health issues, especially for teen boys who want to play high-activity level sports yet need to take caution due to their heightened chance of internal bleeding. Bruises and hemarthrosis (bleeding into a joint cavity) make the disease a visible reminder to youth with hemophilia that they are different, and young adults can find it hard to accept and integrate the illness and treatment in everyday life when they have a job that makes them feel useful in society, and for adults in their early twenties, summer and holidays can be

more stressful than enjoyable due to the possible need for an emergency room visit (Potì et al., 2017).

Further research in the *American Journal of Hematology* shows young adults (ages 18 and up) with hemophilia often struggle with issues regarding their hemophilia as they transition from youth to adulthood, and battling these issues are a part of a mental, physical, and emotional struggle that can lead to being a barrier in hindering access to appropriate care (Quon, et al., 2015). One of these emotional barriers is the necessity to switch care providers. For many young adults with hemophilia, numerous trips to the same HTC for years have "developed quite strong patient-provider relationships and often…are reluctant to leave the pediatric HTC and indeed many continue to be cared for in such a setting even when other options are available" (Young, 2010). Quon et. al (2015) also agrees that there are negative mental consequences on teens with hemophilia who are tackling the transition into adult care, saying the change in personnel "can be felt as abandonment and result in significant stress…and the teen may not feel ready to take on the responsibilities of their complex treatment plans."

An important and defining marker of maturation in society for all young adults is the experience of becoming financially independent from their parents, yet Young (2010) also points out that it can be disheartening to older teenagers looking into a future of independence to carry the knowledge that they may often struggle finding a career that is simultaneously personally and financially rewarding and "does not carry increased risk for bleeding problems." And although youth with hemophilia also desire freedom from being fully dependent on their parents like the majority of all adolescents, much of their emotional support into their young adult years stems from parental assistance with FVIII treatments. The loss of physical parental guidance leaves people with hemophilia in their early twenties at a loss of how to inject FVIII products by

themselves, which can lead to a mindset state of defeat and dismissiveness, and the additional loss of parental insurance can manifest into a burden of anxiety of how to manage treatment costs (Quon et al., 2015). All these worries in the population of youth with hemophilia can accumulate into burdening mental blocks to wanting to pursue the necessary care for their condition.

A solution for the emotional turmoil in adolescents with hemophilia is the start of regular visits to adult hemophilia treatment centers before adulthood so these young adults can be better prepared for the change from pediatric to adult hemophilia care. These individuals could communicate their worries of transition to the HTC staff through meetings or surveys to enhance the focus of what issues need more focus. These discussions should include financial changes that will occur, and perhaps proactive measures and solutions to monetary concerns between patient and provider could be implemented sooner than later. Additionally, a network composed of adults and youth with hemophilia could be created where the elder members could give advice to the younger generation with their shared condition. As for Key Strategies for Improving Employment Among YA-PWH, Quon et al. (2015) suggests a national hemophilia organization increasing patient access to dedicated career counselors with a knowledge of the issues relevant to YA-PWH in order to be an effective way to improve job attainment rates and career satisfaction, all while taking into consideration that the advice should be individualized based on patients' specific interests, skills, goals, and physical limitations.

Care for Hemophilia at Non-Hemophilia Treatment Centers

Because of the spontaneity and severity of the symptoms of hemophilia, people with hemophilia are met with the necessary responsibility of finding care that will promptly and properly assess and treat the issue at hand. Due to the efficacy of the specialized care HTCs

provide, and the consequential improved patient outcomes such as health and quality of life, it is understandable that The World Federation of Hemophilia (WFH) recommends the care of people with hemophilia to be delivered by healthcare professionals at HTCs (de Moerloose, et al., 2020), but patients may still need to attend non-HTC facilities (such as emergency rooms [ERs]) for out-of-hours emergency care of bleeding complications, injuries or factor infusions (Zakieh & Siddiqui, 2017).

In a study run from 2013 to 2015, the Hemophilia Federation of America wanted to discover an estimate of the portion of people with bleeding disorders that received care at non-hemophilia treatment centers. In conclusion, one hundred seventy-seven (22%) participants from 805 eligible surveys were identified as non-HTC people with bleeding disorders, and the most common places for care were the doctor's office (49.4%), ER (16.1%), and hospital outpatient department (12.1%) (Owens et al., 2016). Though the population analyzed was not limited to people with specifically hemophilia as their bleeding disorder diagnosis, and in this research less than a quarter found care at non-HTCs, it is reasonable to want to evaluate the level of care found in healthcare settings that are not specifically designed to treat people with hemophilia. While hardworking healthcare professionals contribute to the wellbeing of their patients to the best of their ability, there can be inevitable shortcomings, whether it be due to preparedness for these patients or knowledge of how to assess an emergency visit, for the treatment of someone with hemophilia that is expressing a secondary hemostatic situation.

In a another study conducted in both the United States and Germany in 2017 to assess the care for people with hemophilia in non-HTCs, hematologists (followed by ER specialists) were the main healthcare providers involved in treating the bleeding cases in question, yet less than 30% of these healthcare professionals/providers (HCPs) in both countries reported to have

protocols for patients with hemophilia (de Moerloose, et al., 2020). Another nearly 30% of United States HCPs in this same study did not know if hospital guidelines existed for hemophilia. In addition, though the time taken for patients with hemophilia to be treated with bypassing agents (such as rFVIII aPCCs) was within an hour at a non-HTC emergency room, the need for approval to use the BPAs and internal process time were proved to be the biggest barriers to immediate care for life threatening hemophilia-related bleeds in the US (de Moerloose, et al., 2020).

In another study conducted from 2019 to 2021 in Canada, patients commented on their own personal experiences with non-HTC care. One participant commented that during emergency room care, they're treated "like…a normal person; somebody that doesn't have hemophilia", and another participant made the statement that because they, as a non-healthcare worker, are not clinically trained, it is difficult to convince healthcare workers (at non-HTCs) that treatment for them is not simply a generic intervention (Arya et al., 2022).

The healthcare professionals, when interviewed, reported that "quicker recognition and better education would improve the outcomes of bleeds in patients with hemophilia and inhibitors…is important and necessary in non-HTC settings" (de Moerloose, et al., 2020). Another idea to assist healthcare settings that do not specialize in hemophilia-associated bleeds is to have around-the-clock availability in HTCs so other HCPs could call and be given advice as to how to assess a hemophilia-related situation, if the need arose. The availability of written guidelines and protocols, and the awareness that such tools exist in non-HTCs are also a necessity to improve care at these facilities, and perhaps the nearest HTCs could work with emergency rooms to create a kind of treatment algorithm.

Though hemophilia is a rare disorder and most likely not a condition HCPs interact with on a day-to-day basis, it is worth noting that it may be *because* of the rarity of the disease that leaves many HCPs in the dark about hemophilia treatment. The purpose of pointing out certain potential risks and limitations of hemophilia care in non-HTCs is not to criticize and belittle the hospital personnel and procedural knowledge surrounding hemophilia, but instead to highlight the evidence that care and awareness for an uncommon condition will vary from one healthcare institution to the next. Though the study had a small sample size and there are not many other reported investigations of obstacles people with hemophilia face when receiving care at non-HTCs other than the ones within the scope of these studies, this does not mean the validity of the concerns surrounding guidance for these individuals should be dismissed. If anything, it shows that the problems are quite real, possible, and ready to be a topic on the forefront of discussions on how healthcare can be improved for not only hemophilia patients, but for other patients who may meet the same types of obstacles to care.

Inadequacies of Care for Women with Hemophilia

Lastly, but certainly not least in importance, is the barrier to hemophilia care that is gravely overlooked: the barriers to hemophilia treatment that are experienced by women. It is notable to remember that with hemophilia, while affecting both males and females alike, the concentration of care is not favored towards women due to the X-linked recessive quality of the inheritance of inherited hemophilia. In an 2018 systematic review that assessed the literature on access to care for women with inherited bleeding disorders, all studies examining bleeding disorders and equity of care in women after the year 2000 were included, yet only a small subset of 526 articles screened focused on women's health, and because an even smaller portion of those findings discussed barriers to hemophilia care in women, it was concluded that "literature

on access to care for women with inherited bleeding disorders…is particularly sparse" (Arya & Sholzberg, 2018).

Though the genetic association for hemophilia for women and men are identical…and women may have FVIII or FIX activity levels equivalent to affected men, exploration surrounding salient issues relating to hemophilia, such as pain management, quality of life, wellbeing, and even psychopathology, have been conducted using an almost exclusively male population (Whitaker et al., 2021; Miller et al., 2021) Out of all patients at hemophilia treatment centers, females were much less likely to have had their factor levels measured, and measuring levels is recommended for all of those undergoing genetic testing, as well as being required for treatment decisions (Miller et al., 2021).

In addition to experiencing the same bleeding symptoms as males, females have the additional risk factors of menstruation, childbearing, and menopause that may require treatment, and an estimated one third of women with hemophilia experience more bleeding than women without hemophilia, especially during menstruation, after dental work, surgery, serious injury or childbirth (Miller et al., 2021). Unfortunately, in addition to the struggle of finding that many HCPs lack awareness of what hemophilia is, there is also the "tendency for hemophilia carriers to encounter dismissive healthcare provider attitudes" (Arya & Sholzberg, 2018).

Because hemophilia is classically an X-linked recessive disease, some healthcare professionals hold to the belief that excessive bleeding symptoms in a female carrier are not possible, which has been repeatedly proven untrue, especially since hemophilia carriers may also experience significant bleeding symptoms because of low factor levels resulting from skewed X‑chromosome inactivation (Punt et al, 2020).

Multiple women with hemophilia have negatively expressed their interactions with healthcare workers, describing their experiences with hemophilia being called into question, or simply not being taken seriously. Though the following study discussed was performed in Canada, yet the same apathetic attitude can be found anywhere where healthcare workers lack empathy and drive to care for their patients. In this investigation, one women cited that she voiced her worries to a HCP that she was bleeding but was told that she was "fine" and to "finish [her] Christmas shopping", as if she "was a wannabe hemophiliac or something" (Arya et al., 2021). Two other women in the same study revealed that even with their pre-existing plans through emergency hemostatic treatment cards (in Canada called Factor First cards), one "wasn't deemed priority enough to receive [an infusion]" even after showing her card, and the other carried her Factor First card "but, a lot of the times, from my experience, it doesn't seem like they either know what it is or care" (Arya et al., 2021).

One of the main reasons to be concerned about the wellness of women with hemophilia, outside of the certainty that they deserve the same amount of validity and attention as their male counterparts, is that the decision to become pregnant or not have children is a factor to be considered by many carriers for hemophilia, and there is an emotional impact of the necessity of having to make such a decision. Overwhelming information during genetic counseling and having to wait for invasive testing procedures leads to stress in hemophilia carriers, and worry and uncertainty of the future for their child during pregnancy, alongside knowing difficulties with hemophilia themselves and in family members, can pose significance influence on their daily lives (Punt et al., 2020).

A possible solution to lessen the unjust treatment for women with hemophilia is the suggestion of changing the nomenclature associated with the classification of women and girls

who have hemophilia. The term “carrier” does not accurately carry the weight of severity of symptoms that this population can and does face, and this leads to the members in this group often being excluded from the conversation and clinical trials surrounding hemophilia. Therefore, there is little understanding about the safety and efficacy and pharmacokinetics of hemophilia therapies in women, and a more inclusive classification system regarding women and girls is urgently needed in hemophilia to enhance awareness, and improve clinical diagnosis, care, education, and research (van Galen et al., 2021). In 2017, there was a successful push from the Scientific and Standardization Committee and the International Society on Thrombosis and Hemostasis to drop the term “carrier” in favor for a new nomenclature that defines women and girls with mild, moderate, and severe hemophilia, according to decreased FVIII or FIX levels, and two additional categories: “symptomatic carrier” or “asymptomatic carrier” were proposed as well to acknowledge that some women with hemophilia with normal levels may have an increased bleeding tendency (van Galen et al., 2021). The only drawback of this nomenclature is the ability to interpret “asymptomatic carrier” as someone with no genuine use for care, so hopefully there will be an emphasis on using “symptomatic carrier” or “hemophilia” when the focus of the interaction is in response to bleeding concerns.

Another solution to help the burden of hemophilia on women is to “ensure good quality, accessible, age appropriate information is given both at diagnosis and at key times during women's lives”, such as puberty, transition to adult services, and family planning, and just like people with other health-related difficulties, women and girls with hemophilia would benefit from “having clear and better access to local services, support groups or counseling at these key time points” (Whitaker et al., 2021).

Finally, similarly to the proposed solution when it comes to care for adolescents with hemophilia and care at non-HTCs, there should still be a strive for better education of healthcare providers about hemophilia-related bleeding symptoms, but in this perspective, the discussions should be focused on symptoms in carriers. There should also be reform and focus "centered on improving HTC policies to formalize appropriate management of carriers, [such as] establishment of Women's Bleeding Disorder Clinics", and in order to begin to rebuild the trust of women with hemophilia to HCPs, the adoption of "clear guidelines for the management of carriers that are widely circulated and accepted throughout the healthcare community" (Renault et al., 2010).

Significance and Discussion

In a world where the advancement of medicine is rapidly evolving, there is the hope among society that cures for diseases (or any type of medical ailment) either exist or are being a priority for the healthcare world, but happens when those solutions currently utilized work appropriately but the problem in question for a particular condition revolves instead around the physical, financial, emotional, and mental barriers to care? The obvious answer is to find resolutions to these as well, but the reality for an already uncommon condition such as hemophilia is that such obstacles are often rarely addressed, so solutions are often simply theoretical and a hope at best for the future. The purpose of this review of the existing literature discussing barriers to care for hemophilia is not to necessarily provide a slew of solutions, but rather to encourage healthcare professionals in all departments to gain an improved understanding that the knowledge of diversely accessible treatment options is essential for their patients.

Conclusion

The author of this thesis is an aspiring medical laboratory scientist, and she does not have hemophilia, so it is reasonable to wonder why the discussion of such a rare disease was deemed a subject of interest worthy of considerable research and thought. However, the status of prevalence of a medical condition shouldn't determine the importance of it, especially to those who will have a direct impact on the health of members in a society where a myriad of different health problems exist. From a medical laboratory standpoint, the extent of laboratorian's involvement with hemophilia may very well not extend the necessary testing to confirm diagnosis of the disorder, but it is crucial as active advocates of other people's physical and mental wellness to remember that on the other side of test results there are real people with aspirations, loved ones, and a life worth living to their fullest extent. Being able to learn about the difficulties of accessible care for just one rare condition has put into perspective for the author of this paper of how needed discussions are for the health of individuals who struggle with underrepresentation, and she hopes that the impact will encourage others, in the healthcare field or not, to come to the same conclusion.

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