Investigating Hemophilia Patients' Experiences and Perceptions of their Health-related well-being in Dhaka City - A Cross-Sectional Study

Thesis paper

[*In the partial fulfillment of the requirements for the degree of Masters of Pharmacy]*

Submitted To

The Department of Pharmacy

Faculty of Allied Health Sciences

Daffodil International University

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APPROVAL

This thesis paper, **"Investigating Hemophilia Patients' Experiences and Perceptions of their Health-related well-being in Dhaka City - A Cross-Sectional Study",** submitted to the Department of Pharmacy, Faculty of Allied Health Sciences, Daffodil International University, has been accepted as satisfactory for the partial fulfillment of the requirements for the degree of Masters of Pharmacy and approved as to its style and contents.

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DECLARATION

I hereby declare that this thesis report, **"Investigating Hemophilia Patients' Experiences and Perceptions of their Health-related well-being in Dhaka City - A Cross-Sectional Study",** is done by me under the supervision Md. Mizanur Rahman Assistant Professor, Department of Pharmacy Faculty of Allied Health Sciences Daffodil International University. I am declaring that this thesis is my original work. I also declare that neither this thesis nor any part therefore has been submitted elsewhere for the award of Master's degree.

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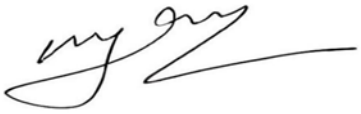
Certificate

This is to certify that the results of the investigation that are embodied in this thesis works are original and have not been submitted before in substance for any degree or diploma of this university. The entire present work submitted as a thesis work for the partial fulfillment of the degree of Masters of Pharmacy.

Supervised By



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My Parents,

The persons who always encourage me in every sphere of my life.

My teacher,

The persons who guided me in this process and the faculties who kept me on track.

Abstract

Background: Hemophilia, a rare bleeding disorder, presents significant challenges to patients' health-related well-being. Understanding patients' experiences and perceptions is crucial for optimizing their care and improving their quality of life.

Methods: A cross-sectional survey was conducted among hemophilia patients in Dhaka City, utilizing structured questionnaires.

Results: As per this survey, 41% of responders reported suffering from hemophilia for 1-5 years. 29% responders have been replied that they have been suffered from hemophilia for 6-10 years. 26% replied that they have been suffered Bleeding of the mouth and gums. 25% responders replied that they have been suffered Bleeding into the joints. 55% retorted that they have been experienced 3-5 times bleeding episode per month. As per this survey most of the responders 60% retorted that their daily activities and mobility has been slightly affected due to bleeding episode. (76%) replied that their family member have been Hemophilia.

Conclusion: This study sheds light on the multifaceted challenges faced by hemophilia patients in Dhaka City concerning their health-related well-being. Further research is warranted to explore additional factors influencing patients' experiences and perceptions, with the ultimate goal of developing comprehensive and patient-centered care strategies.

Keywords: Hemophilia, health-related well-being, patient experiences, perceptions, Dhaka City, cross-sectional study.

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Chapter 1
Introduction

1. Introduction

Hemophilia, a hereditary bleeding disorder, poses significant challenges to individuals affected by it, influencing various aspects of their health-related well-being. In Dhaka City, Bangladesh, where healthcare resources may be limited and awareness about rare disorders like hemophilia could be inadequate, understanding the experiences and perceptions of hemophilia patients regarding their health-related well-being becomes crucial [1]. This cross-sectional study aims to delve into the lived experiences of individuals with hemophilia in Dhaka City, shedding light on their challenges, coping mechanisms, and perceptions of their overall well-being. Hemophilia, characterized by deficiencies in clotting factors, leads to prolonged bleeding episodes even from minor injuries, spontaneous bleeding into muscles and joints, and other complications, significantly impacting the quality of life of affected individuals [2]. In a city like Dhaka, where access to specialized care and comprehensive treatment for hemophilia might be limited, understanding the unique experiences of patients becomes imperative to tailor interventions and support systems effectively. This study employs a cross-sectional design, allowing for the collection of data at a single point in time from a diverse sample of hemophilia patients in Dhaka City. Through structured interviews, surveys, and possibly focus group discussions, the study seeks to explore various dimensions of health-related well-being, including physical, psychological, social, and emotional aspects, as perceived by individuals living with hemophilia [3]. By examining factors such as access to healthcare services, treatment adherence, social support networks, and coping strategies, the study aims to provide a comprehensive understanding of the challenges and resilience mechanisms among hemophilia patients in Dhaka City. Furthermore, this research holds the potential to inform healthcare policies and practices tailored to the specific needs of hemophilia patients in Dhaka City. By identifying gaps in current healthcare provision, highlighting barriers to optimal health-related well-being, and uncovering patients' perspectives on their condition, this study can contribute to the development of targeted interventions aimed at improving the quality of life and overall health outcomes for individuals with hemophilia in this urban setting [4]. In conclusion, this cross-sectional study endeavors to shed light on the experiences and perceptions of health-related well-being among hemophilia patients in Dhaka City. By exploring the multifaceted aspects of

living with hemophilia in this context, the findings of this research can serve as a foundation for implementing effective strategies to enhance the overall well-being and quality of life of individuals affected by this challenging disorder [5].

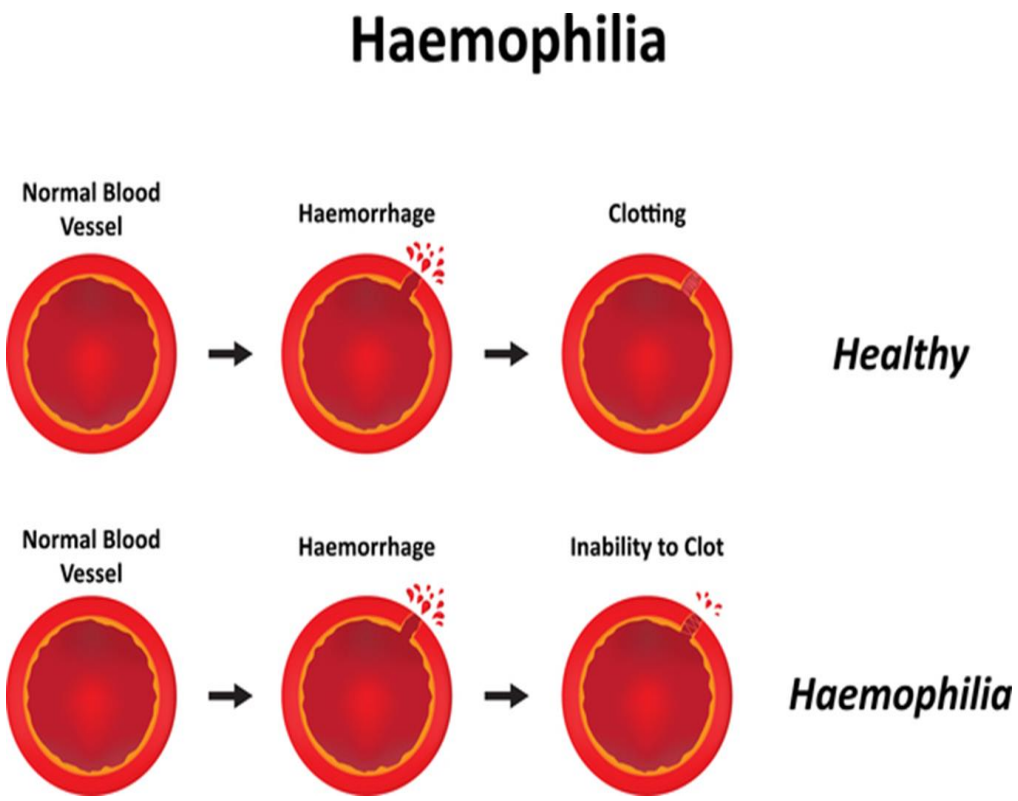


Figure 1: Hemophilia

1.1 Etiology of Hemophilia

Hemophilia is a genetic disorder characterized by a deficiency in certain blood clotting factors, leading to prolonged bleeding episodes. The etiology of hemophilia lies in mutations affecting the genes responsible for the production of clotting factors VIII (hemophilia A) or IX (hemophilia B). Here's a breakdown [6]:

Hemophilia A (Factor VIII deficiency):

Hemophilia A is caused by mutations in the F8 gene, located on the X chromosome. As it's an X-linked recessive disorder, it primarily affects males. Females can be carriers of the gene but are less likely to exhibit symptoms due to the presence of a second X chromosome, which often carries a functional copy of the gene. Mutations in the F8 gene result in reduced

or absent production of Factor VIII, which is essential for blood clotting. This leads to prolonged bleeding episodes and difficulty in forming blood clots [7].

Hemophilia B (Factor IX deficiency):

Hemophilia B, also known as Christmas disease, is caused by mutations in the F9 gene, also located on the X chromosome. Like hemophilia A, it primarily affects males and can be carried by females. Mutations in the F9 gene result in reduced or absent production of Factor IX, another critical clotting factor. Without Factor IX, blood clotting is impaired, leading to prolonged bleeding [8].

Genetic Inheritance:

Both hemophilia A and B follow an X-linked recessive pattern of inheritance. This means that the defective gene is located on the X chromosome. Males inherit one X chromosome from their mothers and one Y chromosome from their fathers. If the X chromosome inherited from the mother carries the defective gene, the son will have hemophilia. Females inherit two X chromosomes, one from each parent. If one X chromosome carries the defective gene, the normal X chromosome may compensate, leading to milder symptoms or no symptoms at all. However, females with one affected X chromosome can still be carriers and pass the gene to their offspring [9].

In summary, hemophilia is primarily caused by mutations in the genes responsible for producing clotting factors VIII (hemophilia A) or IX (hemophilia B). These mutations lead to deficiencies in these clotting factors, resulting in impaired blood clotting and prolonged bleeding. The disorder follows an X-linked recessive pattern of inheritance, predominantly affecting males [10].

1.2 Pathogenesis of Hemophilia

The pathogenesis of hemophilia involves several key molecular and physiological processes:

1. **Genetic Mutation:** Hemophilia is typically inherited as an X-linked recessive disorder. This means the gene responsible for producing clotting factor VIII or IX is located on the

X chromosome. Males have only one X chromosome, so if they inherit a defective gene from their mother, they will develop hemophilia. Females have two X chromosomes, so they can be carriers of the gene mutation but are often asymptomatic due to the presence of a normal copy of the gene on their other X chromosome [11].

2. **Clotting Factor Deficiency:** The genetic mutation in hemophilia leads to a deficiency or dysfunction of clotting factor VIII (hemophilia A) or IX (hemophilia B). These clotting factors are essential for the formation of blood clots to stop bleeding. Without adequate levels of these factors, the blood clotting process is impaired, leading to prolonged bleeding episodes.
3. **Coagulation Cascade:** Clotting factors VIII and IX are key components of the coagulation cascade, a series of enzymatic reactions that culminate in the formation of a stable blood clot. When there is a deficiency of factor VIII or IX, this cascade is disrupted, resulting in ineffective clot formation.
4. **Factor VIII and IX Activation:** Factor VIII and IX are activated through a series of steps involving other clotting factors and cofactors. In the case of factor VIII, it forms a complex with von Willebrand factor (vWF) to stabilize its structure and protect it from degradation. Factor IX is activated by factor XI in the presence of calcium ions and phospholipids on platelet membranes [12].
5. **Platelet Activation and Aggregation:** Platelets play a crucial role in the clotting process by adhering to the site of injury and forming a plug to stop bleeding. In hemophilia, the absence of adequate clotting factor activation leads to impaired platelet activation and aggregation, further contributing to prolonged bleeding.
6. **Fibrin Formation:** Fibrin, a protein formed from fibrinogen, provides structural support to the platelet plug and forms the meshwork of the blood clot. Without sufficient activation of clotting factors VIII and IX, fibrin formation is impaired, resulting in unstable blood clots that are unable to effectively stop bleeding [13].

Overall, the pathogenesis of hemophilia involves a disruption in the coagulation cascade due to deficient or dysfunctional clotting factors VIII or IX, leading to impaired blood clot formation and prolonged bleeding.

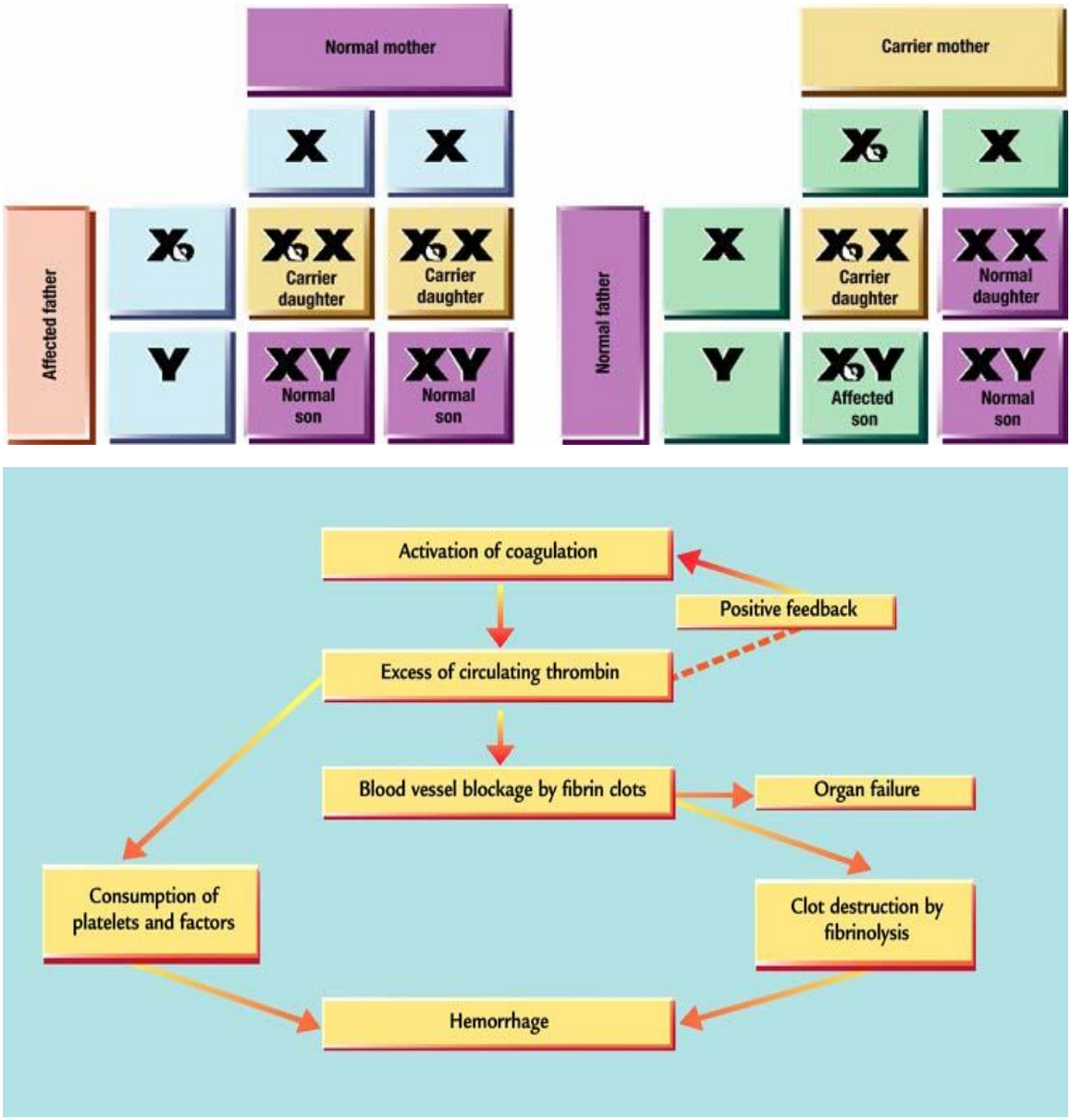


Figure 2: Pathophysiology of Hemophilia

1.3 Diagnosis & Management of Hemophilia

Hemophilia is a rare genetic disorder that impairs the body's ability to control blood clotting. It's caused by a deficiency or absence of specific clotting factors in the blood, most commonly factor VIII (hemophilia A) or factor IX (hemophilia B). Here's an overview of the diagnosis and management of hemophilia:

Diagnosis:

Medical History and Physical Examination: A thorough history of bleeding episodes and a physical examination looking for signs of bleeding disorders are essential [14].

Laboratory Tests:

Clotting Factor Assays: These tests measure the levels of clotting factors VIII and IX in the blood. Low levels indicate hemophilia.

Activated Partial Thromboplastin Time (APTT): Prolonged APTT suggests a deficiency in intrinsic clotting factors, which could indicate hemophilia.

Genetic Testing: Can confirm the specific type of hemophilia and identify carriers [15].

Management:

Replacement Therapy: The mainstay of treatment involves replacing the missing clotting factor through intravenous infusions. This can be either on-demand or prophylactic.

Recombinant Clotting Factors: Purified forms of factor VIII or IX that are genetically engineered.

Plasma-Derived Clotting Factors: Clotting factors derived from human plasma.

Desmopressin (DDAVP) Therapy: In some cases of mild hemophilia A, desmopressin can stimulate the release of stored factor VIII from endothelial cells [16].

Gene Therapy: Emerging treatment option that involves introducing a functional copy of the defective gene into the body to produce the missing clotting factor.

Antifibrinolytic Agents: Medications like tranexamic acid can help stabilize blood clots and reduce bleeding in mucous membranes.

Management of Bleeding Episodes:

R.I.C.E. Therapy: Rest, ice, compression, and elevation can help manage joint bleeds.

Factor Replacement: Infusing the deficient clotting factor to stop bleeding [17].

Physical Therapy: Helps maintain joint flexibility and strength, reducing the risk of joint damage.

Emotional and Psychological Support: Living with a chronic condition like hemophilia can be challenging, so counseling and support groups can be beneficial.

Education and Prevention: Patients and caregivers should be educated about recognizing bleeding symptoms, managing bleeding episodes, and preventing injuries [18].

Complications and Considerations:

Inhibitor Development: Some patients develop inhibitors (antibodies) against the replacement clotting factor, making treatment more challenging.

Joint Damage: Recurrent bleeding into joints can lead to chronic joint damage and arthritis.

Intracranial Hemorrhage: Especially in severe cases, bleeding into the brain can occur, necessitating immediate medical attention [19].

Surgery: Special precautions are required for surgery to prevent excessive bleeding.

Regular Follow-Up: Ongoing monitoring of clotting factor levels, joint health, and overall well-being is crucial for optimal management.

Hemophilia requires lifelong management, but with appropriate treatment and care, individuals with hemophilia can lead relatively normal lives. Close collaboration between patients, caregivers, hematologists, and other healthcare professionals is essential for effective management [20].

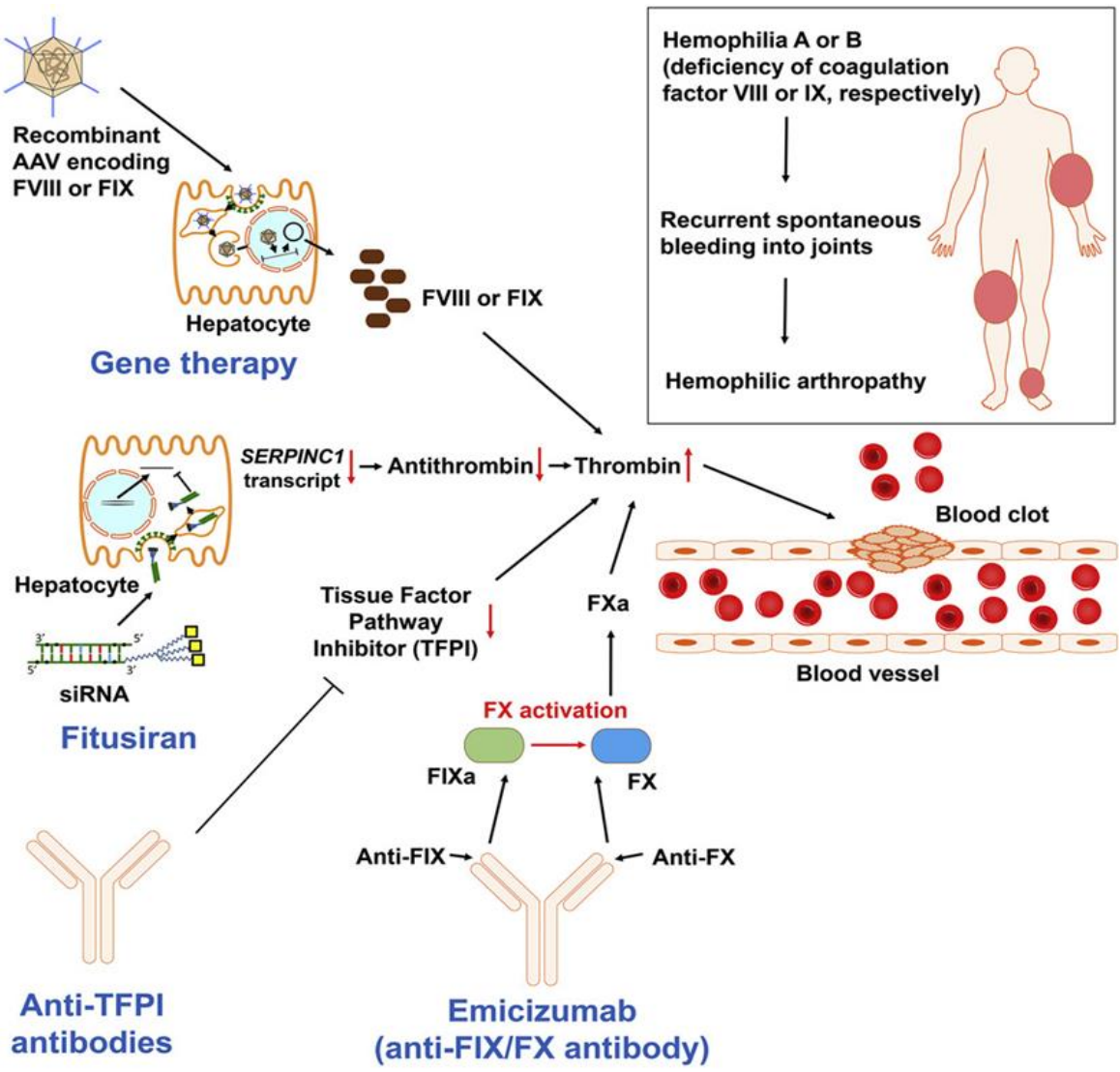


Figure 3: A Molecular Revolution in the Treatment of Hemophilia

Chapter 2

Purpose of the study

2.1 Purpose of the study

General objectives

The primary purpose of this study is to investigate and document the experiences and perceptions of health-related well-being among hemophilia patients residing in Dhaka City.

Specific objectives

- To understand the overall quality of life of hemophilia patients, focusing on physical, psychological, and social aspects.
- To identify specific health challenges and issues faced by hemophilia patient.
- To evaluate the accessibility, effectiveness, and satisfaction.
- This study aims to contribute to the existing body of knowledge on hemophilia.

Chapter 3
Literature Review

3.1 A Qualitative Study Exploring the Experiences and Perceptions of Patients with Hemophilia Regarding Their Health-Related Well-Being, in Salamanca

A persistent X-linked condition called hemophilia is caused by a deficiency in factors VIII or IX, which are essential for blood clotting. Hemophiliacs frequently experience unique psychosocial issues related to accepting, coping with, treating, and managing their illness as well as their family and social interactions, which are frequently impacted by these situations. This study set out to investigate the effects of hemophilia on the lives of individuals with the disorder or those close to them in a particular area of Spain. A review of the literature of qualitative hemophilia research was done, and organized interviews were constructed and performed utilizing the World Organization of Hemophilia and Osorio-Guzmán et al. research as an outline. The findings demonstrated that three major themes arose from the data: the effects of hemophilia on a daily basis, ambiguity surrounding the illness, the function of connections, and institutional assistance. The findings demonstrate how significantly the illness affects their lives in terms of job, family, leisure, and personal surroundings. The primary finding is that hemophilia negatively affects patients', families', and caregivers' daily life [21].

3.2 The management of hemophilia in elderly patients

After the increasing rate of deaths observed during the 1980s due to human immunodeficiency virus (HIV) infection, the health-related quality of life and life expectancy of persons with hemophilia have improved, mainly due to the progresses of replacement therapy and antiviral drugs and to the improvement of the global comprehensive care provided by specialized centers. As a consequence, an increasing number of hemophiliacs have reached an older age and nowadays physicians in hemophilia centers find that they must handle age-related clinical problems never previously observed in this population. The management of elderly persons with congenital hemophilia is discussed in the first part of this review. The second part describes the general aspects of acquired hemophilia due to anti-factor VIII autoantibodies, focusing on the clinical management of elderly patients, one of the groups most frequently affected by this acquired bleeding disorder [22].

3.3 Assessing health-related quality-of-life in patients with severe hemophilia A and B

Although severe hemophilia is a continuously painful and incapacitating circumstance, therapy may slow the disease's progression and, as a result, improve health-related quality of life (HR-QoL). 99 patients with serious hemophilia A and B were given questionnaires from MOS Short Form 36 (SF-36) and EuroQol (EQ-5D) to evaluate HR-QoL. Considered were the correlations among the two questionnaire replies, age, HIV status, previous experience with orthopedics, and the quantity of bleeding episodes patients had endured in the preceding year. Additionally, scores from both surveys were contrasted with relevant UK normative data. Based on 70 and 71 SF-36 and EuroQol surveys, correspondingly, the final analysis was conducted. Apart from age, no clinical characteristic was shown to be a statistically significant indicator of quality of life linked to health (HR-QoL). Regardless of variations in age, the results from both questionnaires made it abundantly evident that these patients with serious hemophilia have far lower HR-QoL than the general population [23].

Chapter 4
Methodology

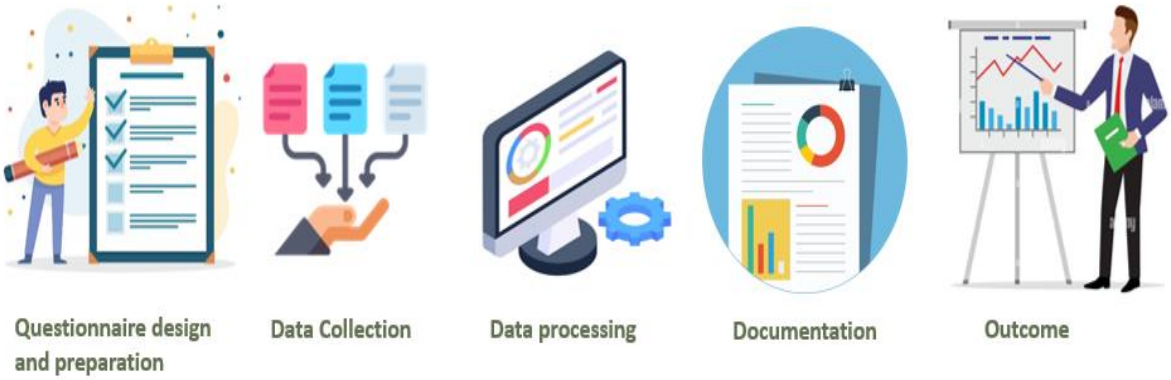
4. Methodology

Descriptive cross-sectional study was conducted to determine patients Experiences and Perceptions of their Health-related well-being in Dhaka City.

- The study period was January 2024 to April 2024.

4.1 Data Collection:

- Develop a structured questionnaire that covers demographics, awareness about Hemophilia, prevalence, and management practices.
- Pre-test the questionnaire to identify and address any issues.
- Employ trained interviewers for face-to-face or telephone interviews, depending on resources.
- Translate the questionnaire into Bengali for better understanding by the local population. Train a team of data collectors on the questionnaire and ethical considerations.
- Conduct face-to-face interviews, phone interviews, or online surveys, depending on the accessibility of the respondents. Ensure informed consent and data confidentiality.
- Some important data has been collected by reviewed number of related article paper from different website like Google scholar, research gate and PubMed.



4.2 Data Variables:

- Collect demographic information, including age, gender, educational background, occupation, and residence.
- Gather evidence associated to Understanding Prevalence & Management of hemophilia, such as the presence of symptoms, family history, triggers, severity, and healthcare utilization.
- Explore lifestyle factors, including smoking habits, dietary patterns, and physical activity.
- Assess environmental factors, such as air quality, housing conditions, and proximity to potential allergens.

4.3 Sample size

The test had 13 short-answer questions and took roughly four to five minutes to finish.

- I have tried my best to collect all data from different profession people for gathering different types of information.
- The examination is led by a questionnaires oriented survey, around 200 populations was being responded for this assessments.

4.4 Data analysis strategy

Data analysis is the methodical application of statistical and/or logical tools for describing and illustrating, condensing and summarizing, and evaluating data. Use appropriate statistical software to analyze the collected data. Employ descriptive statistics to summarize demographic and prevalence data.

Chapter 5

Results & discussion

5.1 Age of Responders

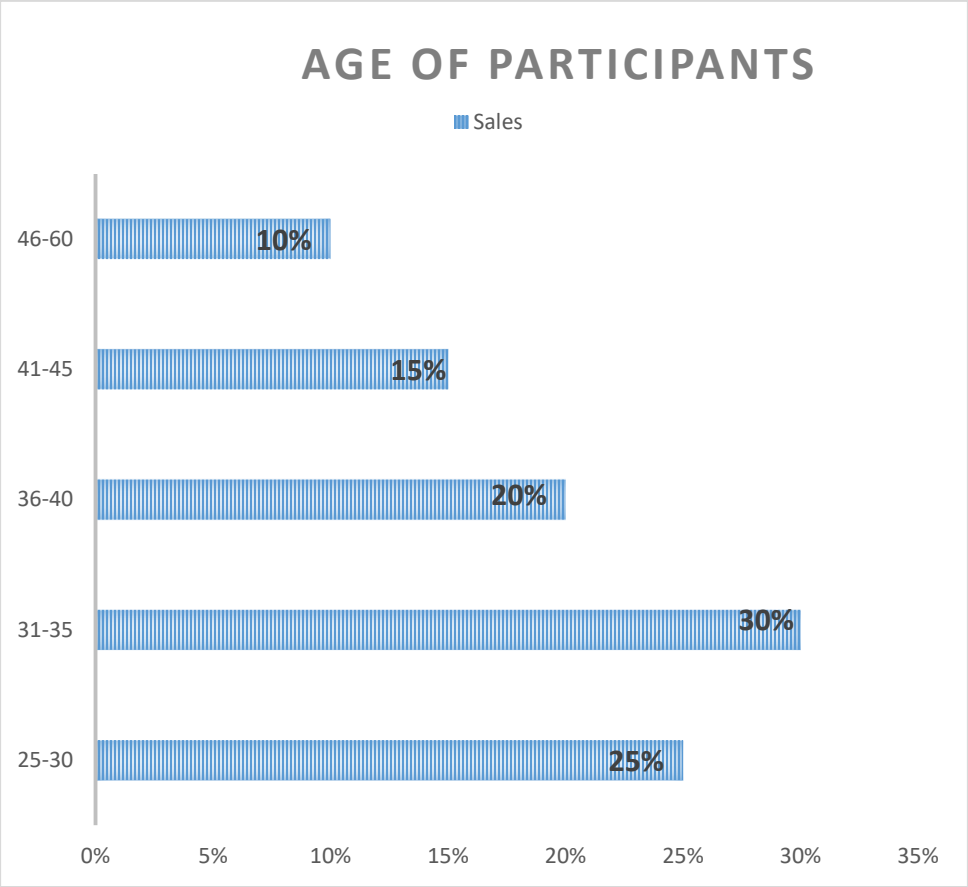


Figure 4: Age of Responders

Discussion: There were 200 participants in all for this investigation. Respondents ranging in age from 25 to 60 were between them. Here, the largest age group of participants (30%) was between 31 and 35 years old. Participants aged 25 to 30 made up 25% of the group, followed by those aged 36 to 40 (20%), 41 to 45 (15%), and 46 to 60 (10%).

5.2 Gender of Participants

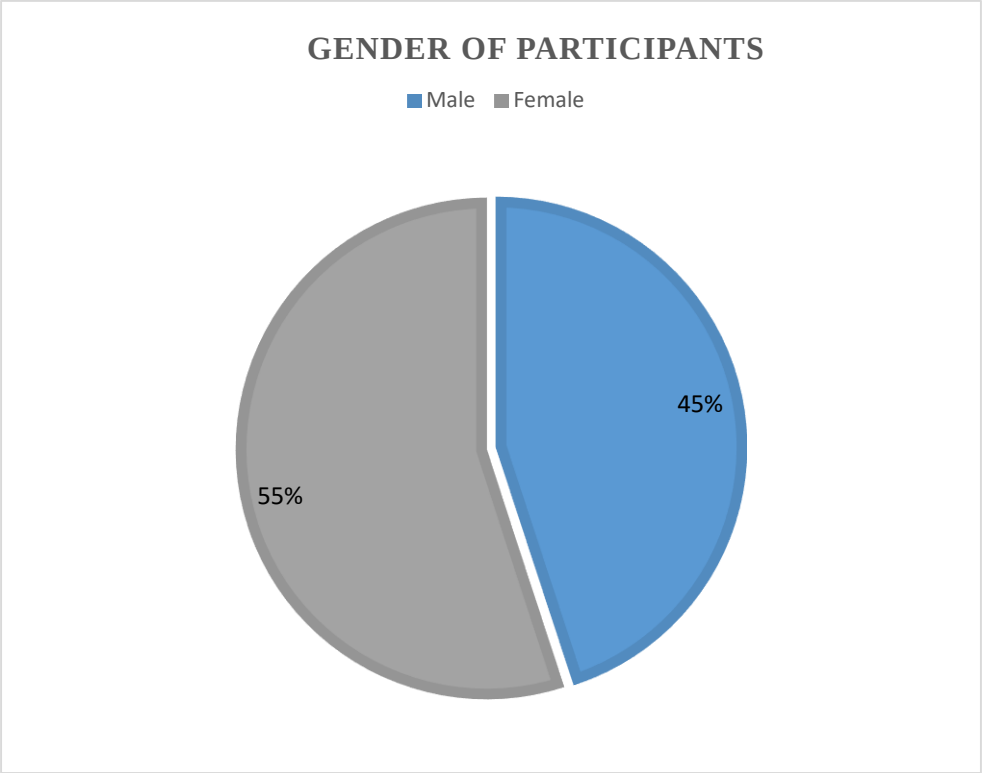


Figure 5: Gender of Participants

Discussion: In the Western world, psoriasis is a prevalent skin ailment that affects 2-4% of people. While the frequency rises more quickly in younger female patients as they get older. The diverse gender representation in this inspection, here 45% male and 55% female members.

5.3 Professional Status of Responders

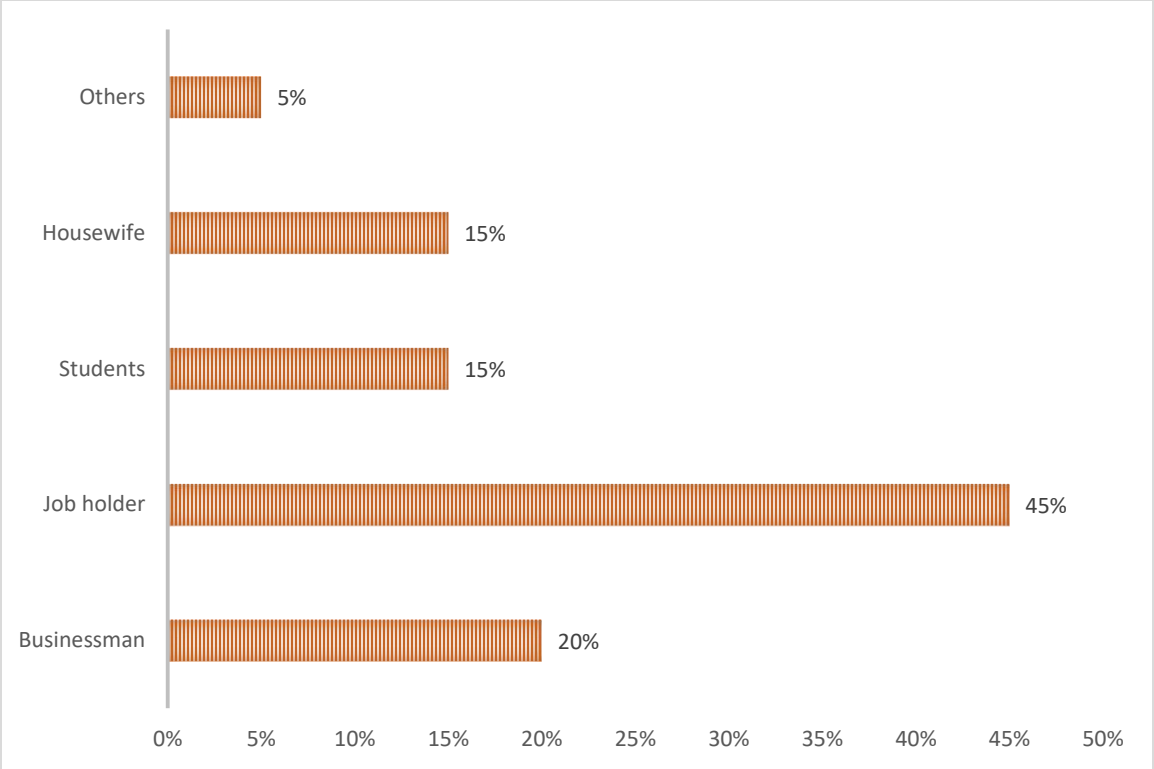


Figure 6: Professional Status of Responders

Discussion: Different professional personnel has been chosen for this survey. Conferring to figure 6 most of the participants are Job holder (45%), also 20% are businessman.

5.4 Familiar with hemophilia

Q: How familiar are you with hemophilia?

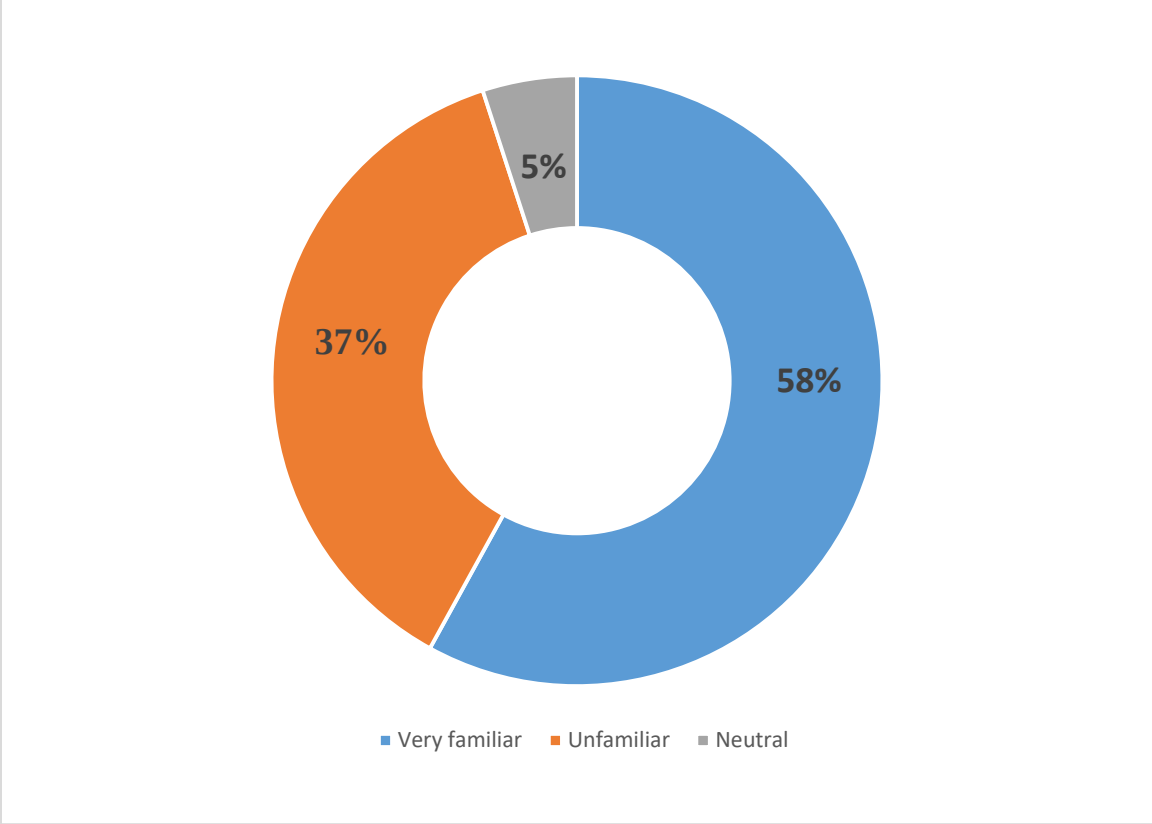


Figure 7: Familiar with hemophilia

Discussion: Blood clotting is disrupted in hemophilia, a hereditary bleeding illness. It results from one of the clotting proteins in the blood being absent or present in less than normal amounts. As per this investigation (58%) responders replied that they have been very familiar about hemophilia. 37% of respondents retorted that they have been unfamiliar about hemophilia.

5.5 Affected ratio of Hemophilia

Q: Have you ever been affected by Hemophilia?

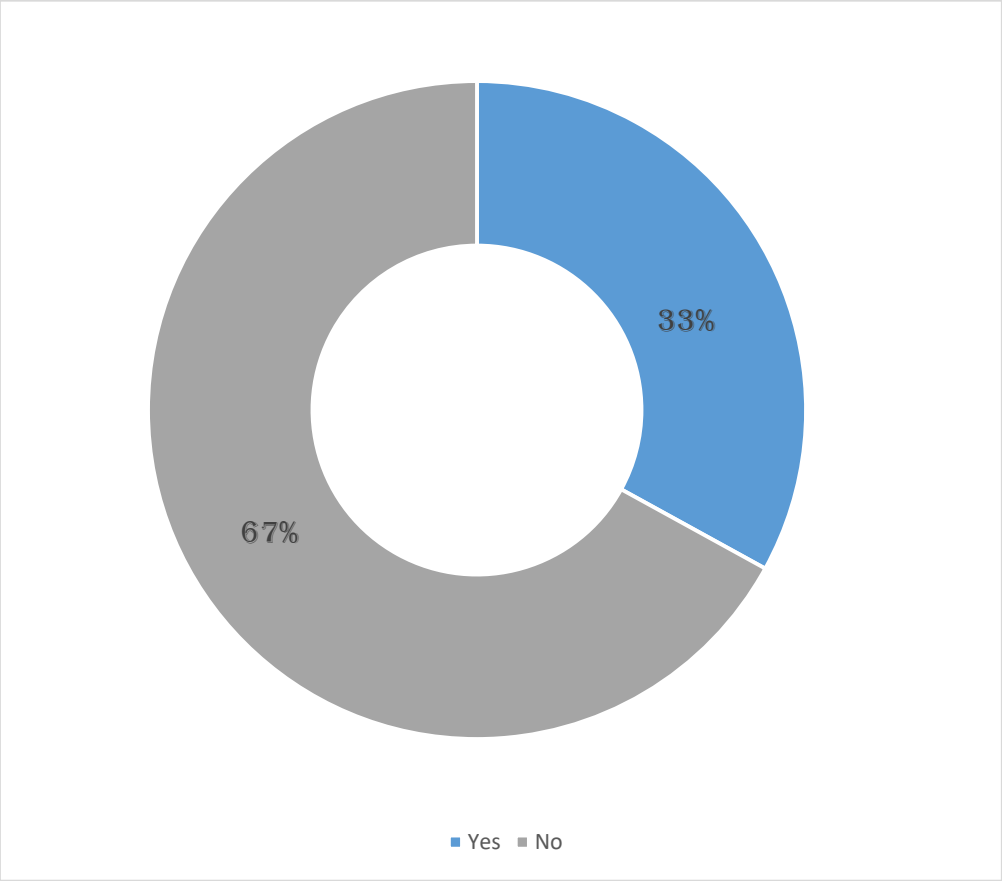


Figure 8: Affected ratio of Hemophilia

Discussion: As per this survey most of the responders (67%) haven't been affected Hemophilia. 33% replied that they have been affected with Hemophilia.

5.6 Duration of illness

Q: How long have you been diagnosed with hemophilia?

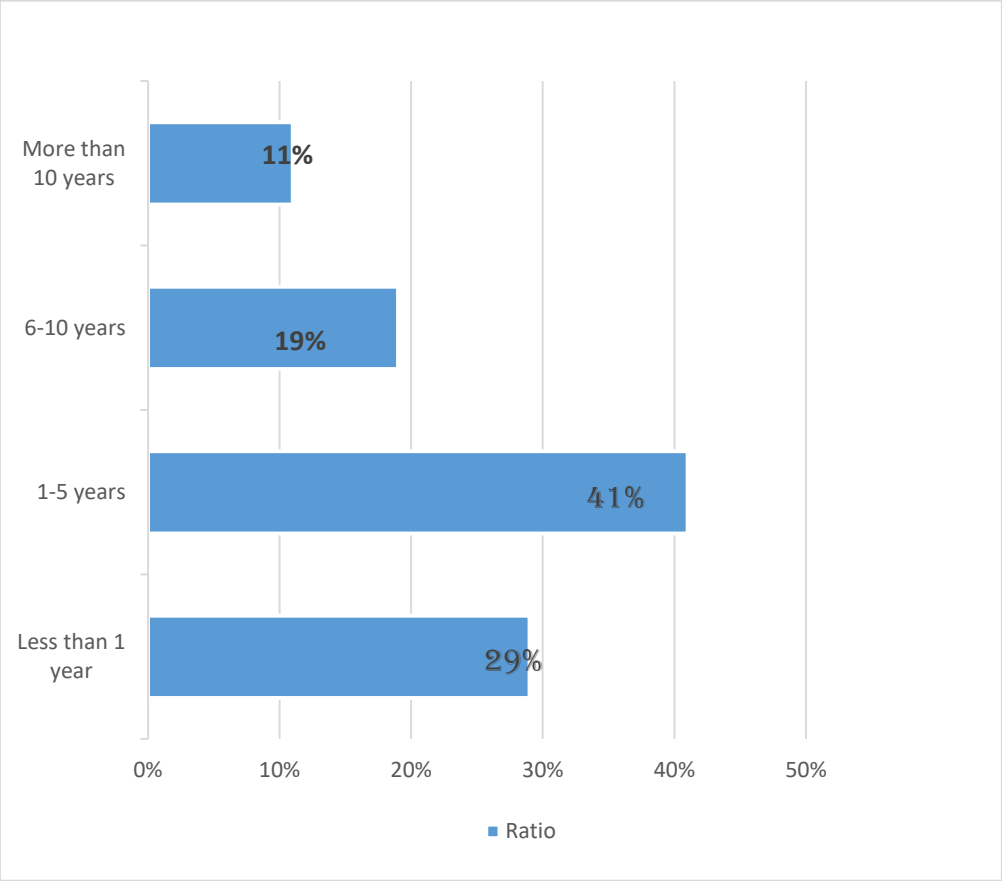


Figure 9: Duration of illness

Discussion: As per this survey, 41% of responders reported suffering from hemophilia for 1-5 years. 29% responders have been replied that they have been suffered from hemophilia for 6-10 years & 19% responders replied that they have been suffered from hemophilia for less than one year.

5.7 Symptoms of Hemophilia

Q: If yes, what kind of symptoms have you experienced?

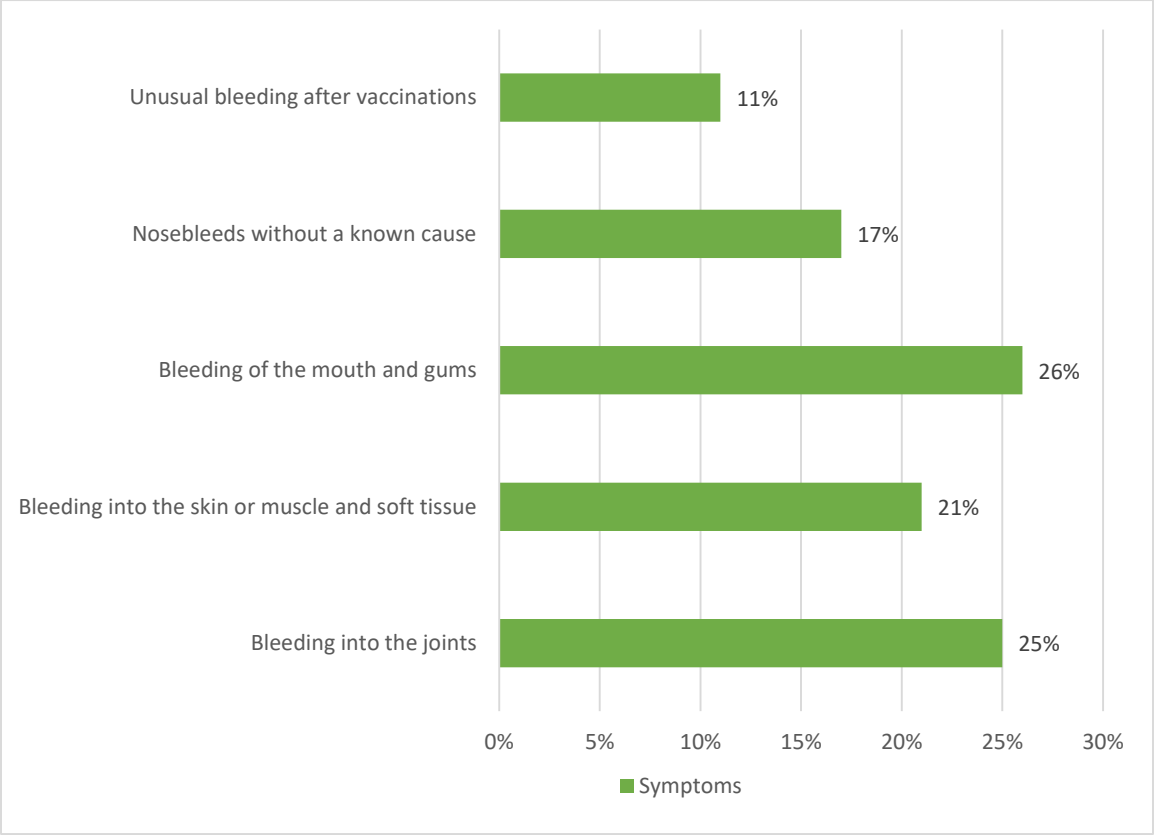


Figure 10: Symptoms of Hemophilia

Discussion: Many of the afflicted patients experienced a variety of symptoms. Conferring to the investigation among the contributors 26% replied that they have been suffered Bleeding of the mouth and gums. 25% responders replied that they have been suffered Bleeding into the joints, 21% responders replied that they have been suffered Bleeding into the skin (which is bruising) or muscle and soft tissue causing a build-up of blood in the area (called a hematoma) & 17% responders replied that they have been suffered Nosebleeds without a known cause.

5.8 Frequency of bleeding per month

Q: "On average, how many times do you experience bleeding episodes per month?"

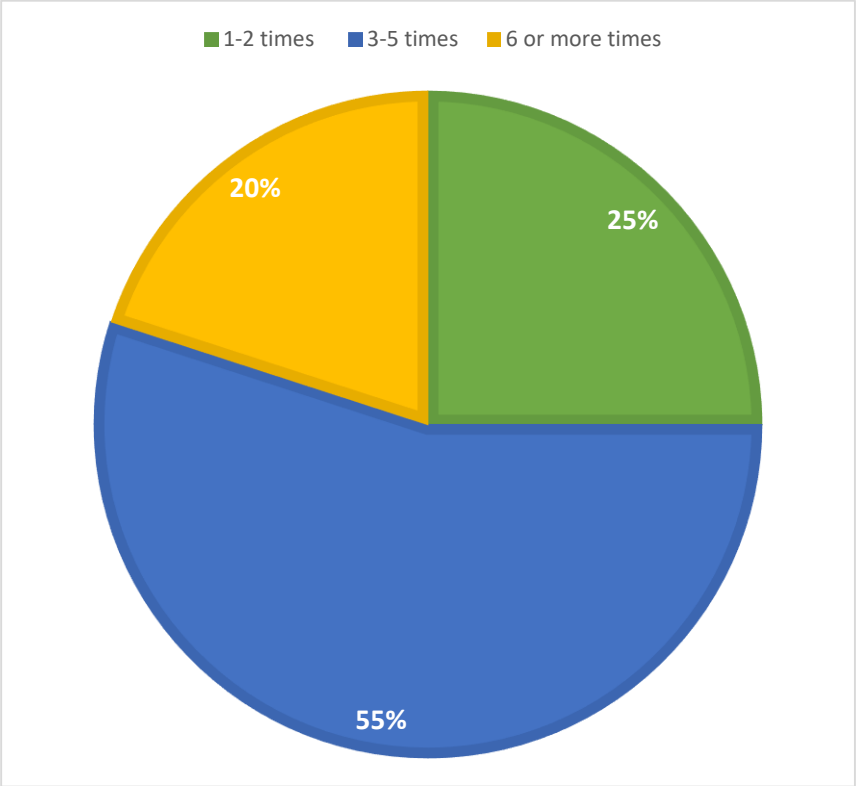


Figure 11: Frequency of bleeding per month

Discussion: The number of bleeding events that occur during a specific time. As per this survey most of the participants 55% retorted that they have been experienced 3-5 times bleeding episode per month. 25% responders replied that they have been experienced 1-2 times bleeding episode per month & 20% participants retorted that they have been experienced 6 or more times bleeding episode per month.

5.9 Affected daily activities and mobility due to bleeding episodes

Q: How do bleeding episodes affect your daily activities and mobility?

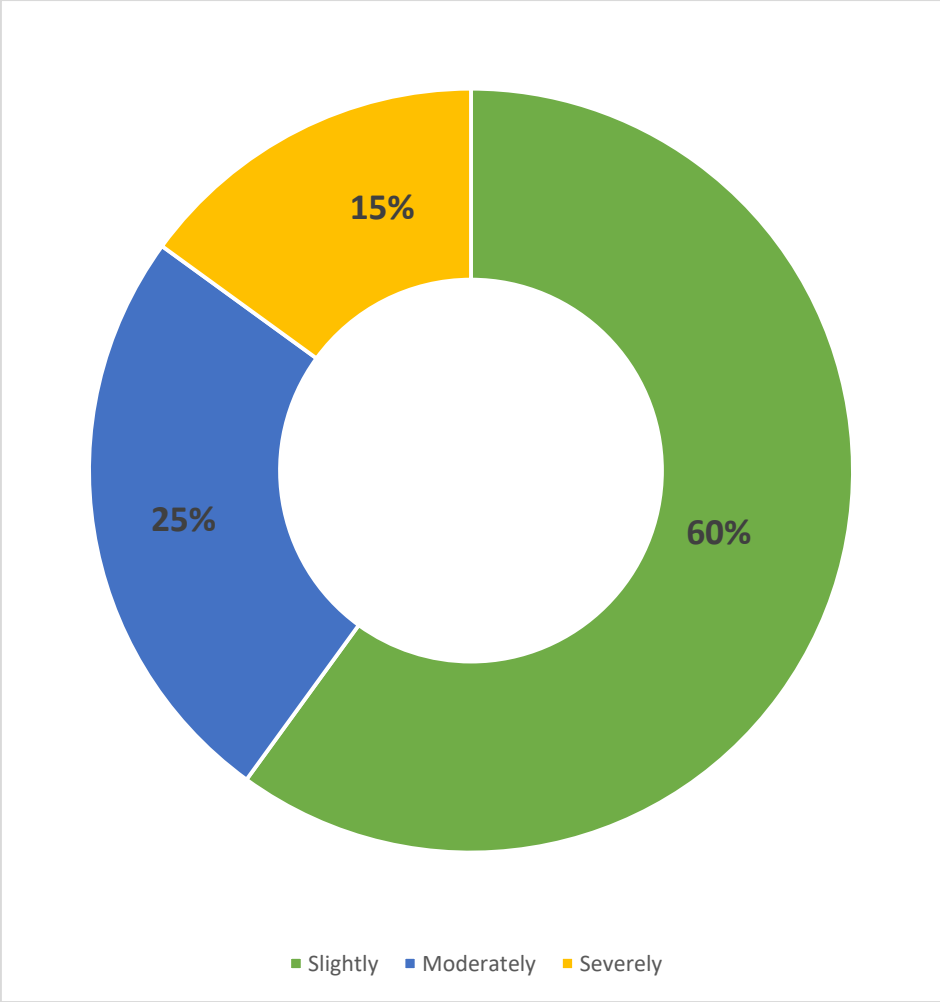


Figure 12: Affected daily activities and mobility due to bleeding episodes

Conversation: Individuals with bleeding disorders wish to live regular lives unaffected by their illness, which is within grasp. As per this survey most of the responders 60% retorted that their daily activities and mobility has been slightly affected due to bleeding episode. 25% retorted that their daily activities and mobility has been moderately affected due to bleeding episode.

5.10 Agree with the following statement

Q: “The best way to treat hemophilia is to replace the missing blood clotting factor so that the blood can clot properly” Do you agree with the following statement?

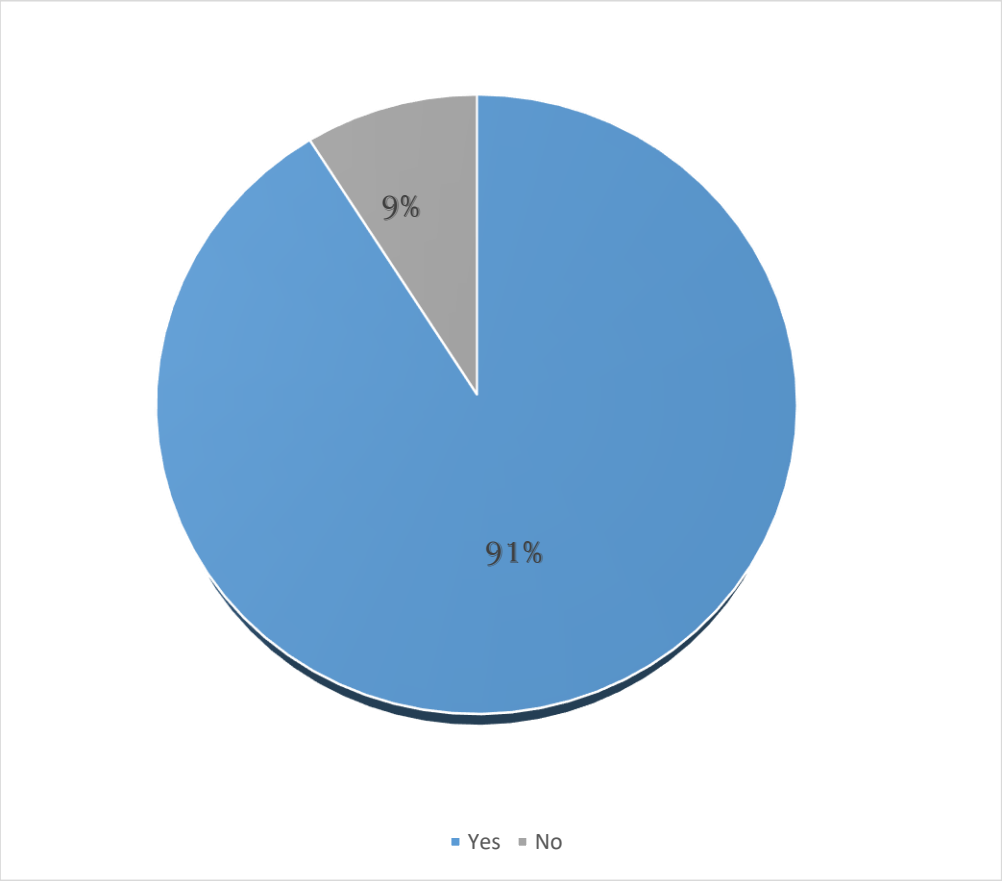


Figure 13: Agree with the following statement

Discussion: As per this investigation, most of the participants (91%) retorted that they have been greed with **“The best way to treat hemophilia is to replace the missing blood clotting factor so that the blood can clot properly”** this statement.

5.11 Agree with the following statement

Q: “Neutralizing antibodies, against the factor is the biggest issue. It has been created challenges with current hemophilia treatment” Do you agree with the following statement?

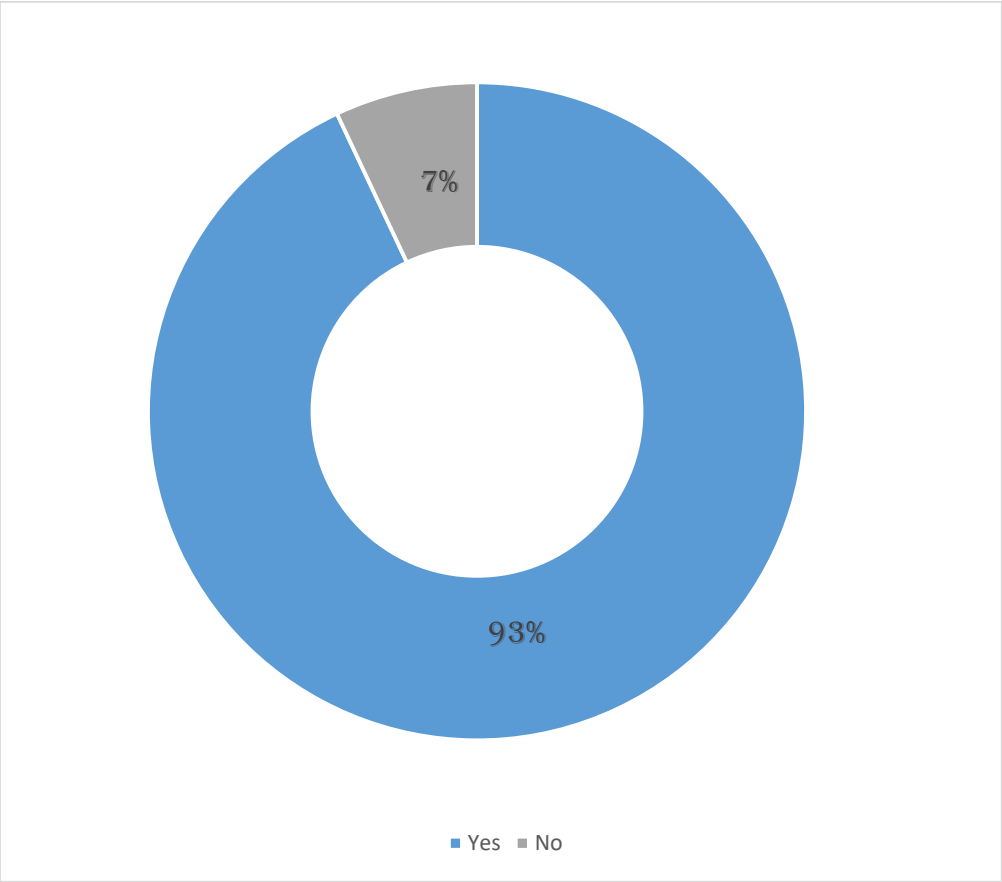


Figure 14: Agree with the following statement

Discussion: As per this investigation, most of the participants (93%) retorted that they have been greed with “Neutralizing antibodies, against the factor is the biggest issue. It has been created challenges with current hemophilia treatment” this statement.

5.12 Family history

Q: Do you have a family history (like father, mother and siblings) who has or had Hemophilia?

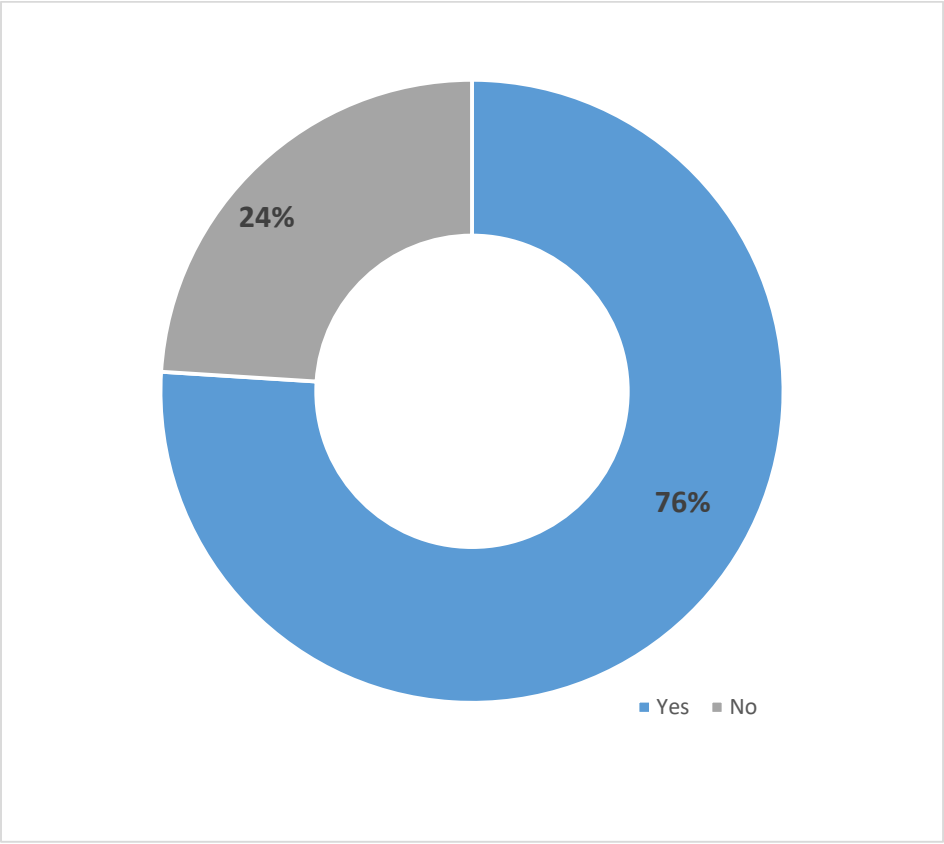


Figure 15: Family history

Discussion: Hemophilia is a congenital disorder that runs in families. Hemophilia is brought about by a mutation or change in the gene that produces factor VIII (8) or factor IX (9). In this investigation most of the responders (76%) replied that their family member have been Hemophilia.

5.13 Participated in any hemophilia support groups or community events

Q: Have you ever participated in any hemophilia support groups or community events?

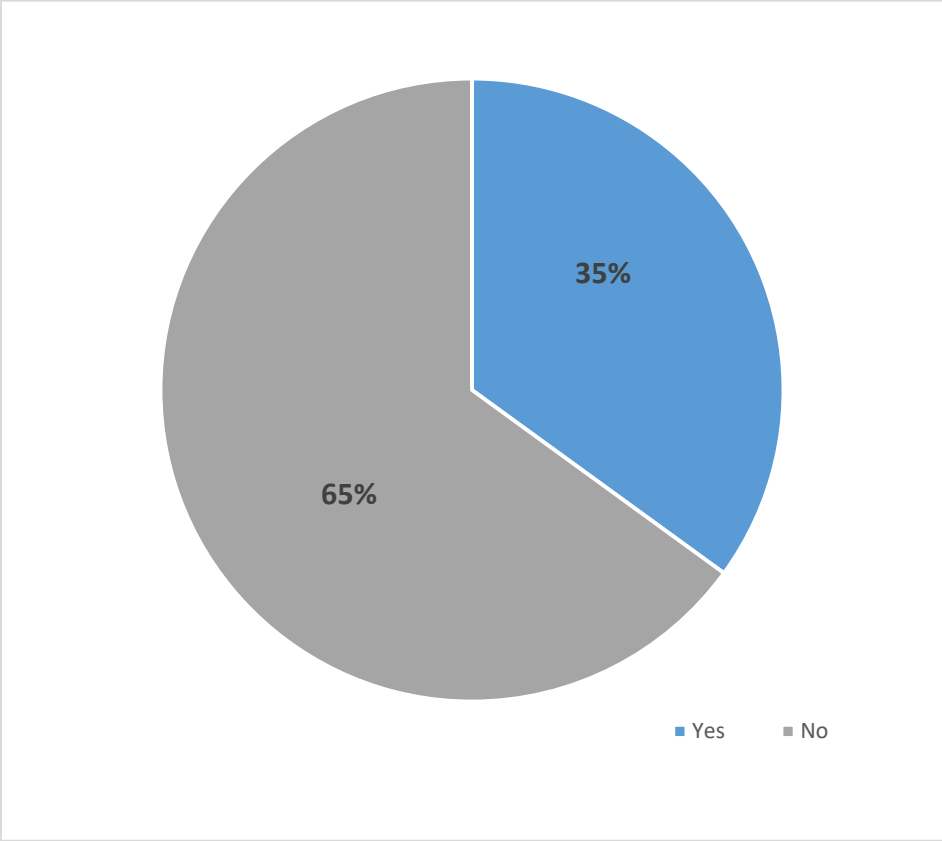


Figure 16: Participated in any hemophilia support groups or community events

As per this investigation, 35% responders replied that they have been participated in hemophilia support or community events.

Chapter 6
Conclusion

6. Conclusion

In conclusion, this cross-sectional study sheds light on the experiences and perceptions of hemophilia patients regarding their health-related well-being in Dhaka City. Through comprehensive data collection and analysis, key insights have been gleaned into the challenges, limitations, and coping mechanisms adopted by individuals living with hemophilia in this urban setting. Overall, this research contributes to a deeper understanding of the lived experiences of hemophilia patients in an urban context and underscores the importance of holistic approaches to address their health-related well-being.

Chapter 7
Reference

Reference

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