

ASSESSMENT OF PREVALENCE AND PRESCRIPTION TRENDS OF PATIENTS WITH HEMOPHILIC DISORDER

Aklima Akter* Farhana Israt Jahan Nahid Rahman Esha
¹Daffodil International University, Shukrabad, Dhaka, Bangladesh

* aklima.ju@gmail.com, jifarhana@daffodilvarsity.edu.bd, rahman.asha11@gmail.com,

Abstract

Hemophilia is a congenital rare disorder commonly found in male rather than female. This rare disorder can affect approximately 1 in 10000 people in worldwide. It is an inherited genetic disorder where patients face shortage of blood clotting factors. Affected patients are taken injection factor 8 and factor 9 and external blood from any source to manage this disease. No vaccination is available in worldwide to remove this disorder. To understand the current practice and knowledge of Hemophilia patients in Bangladesh, this study is done at Bangladesh medical college hospital, Bangabandhu Sheikh Mujib medical university Hospital, Hemophillia society (Kolabagan) among 70 patients, collection of their concurrent prescription and analyzed in different ways such as gender, etiology, common causes, symptoms, medication pattern, doctor visiting frequency, knowledge about the disease etc. and found that it is most commonly found in male than female (98.57% of male), people in urban area is at high risk for this disease (48.57% of patients in Dhaka), it can occur in patients mainly by genetically (81.81% of patients inheritably for Type A & B), joint knee pain is the major symptoms found in affected patients (34.28% of knee pain symptom), injection (72.72% of patients in Type A & B) and fresh plasma (27.28% of patients in Type A & B) are two major treatment options for affected people, patients have to face daily doctor visit (62.85% of patients), major percentage of patients have no knowledge about this disease (74.28% of patients). It is explicit that nowadays hemophilia is getting popular in Bangladesh and people make acquaintance with this disorder.

Keywords: Hemophilia¹, Hemophilia², male³, blood⁴, clotting factor⁵, plasma⁶

Introduction

Hematology is the practice of blood. There are many kinds of bleeding disorder. Hemophilia is a rare inherited bleeding disorder mainly seen in the male. It is also called a royal disorder because this disease was passed from Queen Victoria, who became Queen of England in 1837.[1] Hemophilia is occurred by the lack of clotting factor VIII & IX in the body as a result blood does take a longer time to clot than normal clotting time. The prevalence of hemophilia is estimated to be about 1:10,000 birth and that of the severe form of the disease to be about 6% per 1,00,000 population worldwide. Three types of hemophilia are seen that is A, B, C. Specific dominance rates from the six scrutiny states to the U.S. inhabitants resulted in a predictable national, inhabitants of 13,320 personal belongings of hemophilia A and 3,640 personal belongings of B. [2] The lack of precise country-specific dominance data has unnatural planning efforts for the handling. Hemophilia A is a type owed to factor VIII deficit. The prevalence of this type is about 1% worldwide. Hemophilia B is another type due to factor IX insufficiency. It is also called Christmas disease. Another type of hemophilia is called acquired hemophilia and it can occur by lacking factor XI. It's very rare in Bangladesh. The rate of this type occurs less than 1% worldwide. In the survey study, hemophilia type A and type B is taken into consideration. Type of A is very regular in Bangladesh on the other hand type of B is very uncommon. Though it is a genetic disorder, it can pass from mother to fetus and mother is a media of this disorder. It occurred by the defective of X chromosome hemophilia passed from mother to fetus. A hemophilic mother who acts as a carrier has half of the amount of factor VIII & IX than normal. In percentage, about 10 percent of carrier females have less than half the normal amount of one of these coagulation factors which can create risks of abnormal bleeding. [3] In hemophilia A and B type, males are found affected than female. Approximately 1 in 5,000 males are born with Hemophilia A, and 1 in 30,000 males is born with Hemophilia B. [4] But in hemophilia type C, male and female are affected equally. So hemophilia is not related to a single factor, sex chromosome. Hemophilia is diagnosed all the way through a blood

assessment. Physicians may resolve confiscates a little taster of blood commencing vein and assess the quantity of clotting factor in attendance. Only about 25% of the estimated 400000 people with hemophilia global obtain sufficient management. [5] Hemophilia is a rare disorder that is complex to diagnose and to manage. Some evidence-based guidelines may help in proper management and diagnosis of hemophilia, as well as the maintenance of other hemophilia induced complications. [6] Hemophilia A is treated by the prescription hormone and the name of the hormone is desmopressin. This hormone is used by putting an injection into a vein. This injection stimulates the factor VIII. Hemophilia B is treated by transfusion of blood from the donor. Sometimes, the factors may be given in the artificial form. These are called "recombinant clotting factors." Approach to reducing complications by hemophilia C is plasma combination. Usual treatments used by patients either take an injection or external blood to manage this disorder. Hemophilia can be identified by blood screening examination, absolute blood calculation, activated partial thromboplastin time, prothrombin time (PT) examination, fibrinogen examination etc. Hemophilic patients would identify some sign and symptoms like frequent nosebleeds, joint pain, bleeding gum. Other types of symptoms are: vomiting constantly, pain in the neck. To make awareness among people and to provide care and information to affected people, in 1963, a person with hemophilia disorder named Frank Schnabel established the World Federation of Hemophilia. On World Hemophilia Day, April 17, 2003, the WFH launched the Global Alliance for Progress (GAP) in hemophilia which aims to double the number of people with hemophilia diagnosed and receiving treatment in up to 40 developing countries over a 10-year period. [7] Along with the world, in Bangladesh, we have to celebrate the hemophilia day so that we have to people gather knowledge and make awareness to cure and controlling this rare inherited disorder.

Methods

A cross-sectional study is used in this survey practice. This is the innovative technique that determines the opinions of the targeted people by the collecting of information from the targeted

population by asking various kind of question. It is the co-relational way to combine or gather opinion or attitude by the people. The practice was conducted from February 2018 to June 2018. This study is conducted by close observation of patients, questionnaire, and evaluation of the prescription. In this practice, about 70 patients were elected from Bangladesh medical college hospital, Bangabandhu seikh mujib medical university Hospital, Hemophilia society (kolabagan). The elected participants were Bangladeshi in origin and patients' age was varied from 2-70 years. In this practice indiscriminately sampling was mimicked. After collecting, the entire datum was confined and analyzed with the help of Microsoft Excel.

Results

In this survey practice on 70 patients, variations of results are found on many criteria such as age, area, gender, medication pattern, mostly prescribed medicine by doctors, patients' doctor visiting frequency etc.

According to the table, the prevalence of gender of patients (Table 1), it can be seen that male people are affected highly in this diseases and in percentage, 98.57% of patients are found male and only 1.42% of patients are female.

According to the table, the age of patients (Table 2), people of 0-18 age range are mostly affected and least affected people are found in 45-60 age group. Results that are shown in table 2 that patients in 0-18 age range 59.93% are affected (Type A) and 66.66% of patients (Type B) are suffered from this disease. In the age group of the 18-25 range, results are slightly varied for Type A 16.94% and Type B 8.33%. For the age group of the 25-45 range, 18.64% patients are found from Type A and 16.66% from Type B. Among elderly patients, 8.47% are found in Type A and 0% is found in Type B.

According to the figure of the area of patients (Figure no. 1), It can be said that Dhaka is highly affected in this diseases, also available in Pabna, Dinajpur, Rangpur etc. In percentage, 49% of patients are from Dhaka district, second highest affected area found is Pabna (19% patients), third largest groups are from Dinajpur (17% patients), in percentage another district are found as Rangpur (11% patients) and Sylhet (4% patients).

According to the figure of causes of hemophilia (Figure no. 2), it is found that mainly this disease occurred by genetically. Almost 80-90% cases patients inherently suffered from this disease. In the case of Type A, the reason for this disease is found genetically about 89.83% patients and for Type B it is found about 81.81% patients. The unknown cause is found 10.17% patients for Type a and 18.18% patients for Type B.

According to the figure of symptoms of patients (Figure no. 3), it is seen that knee joint swelling (34.28% patients) is very common symptoms among all patients. Similarly, shoulder pain (21.42%), blood in the urine (20%), bleeding gum (14.28%) are seen among patients. Very less common symptoms are found as nosebleed (10% patients).

According to the table of medication pattern of patients (Table no. 3), it is observed that anti hemophilic injection is very regular inpatients and patients also take Fresh frozen plasma in some cases. About 70-80% of patients regularly have taken an anti-hemophilic injection and 20-28% patients have taken fresh frozen plasma.

Figure no. 4 has shown that anti-hemophilic injections taking frequency of both Type A and Type B patients are almost similar. Daily injection taking frequency of Type A patients found 70.38% whereas Type B patients are found 66.67%. Other results are found in a weakly manner treatment, where Type A patients are found 22.22% and the similar result is found for Type b patients as 22.22%. In a monthly manner treatment, Type A patients are found 7.04% and Type B patients are found 11.11%.

According to the figure of freshly frozen plasma frequency of patients (Figure no. 5), it can be seen that hemophilic patient need also fresh frozen plasma. Daily fresh frozen plasma taking frequency is found similar in Type A and Type B patients are about 66.67%. Slight differences have shown in results of weekly and monthly treatment. In a weakly manner, the result found in Type A is about 25% and in Type B patients is about 33.33%. Again, in a monthly manner, patients' percentage for Type A is found 8.33% and for Type B is found 0%.

From the Table no. 4, it is found that people awareness are very low about this bleeding disorder; only 25.71% of patients are acquaintance

with hemophilia disorder and most of them have no knowledge (74.28%).

According to the figure of others medication pattern of patients (Figure no. 6), It is found that Napadol (Tab.) is a very common medicine which is taken by the hemophilic patients and it is about 39%. Second most prescribed medicines are found Traxyl (Inj.) (26%) and Ace (21%) & Traxyl (Cap.) (14%) is found accordingly.

Discussion

Among all type of study, prescription based study is mostly a professional study whereas physicians reflection is found directly about patients and it is also helpful to assess and evaluate physicians prescription pattern for disease management. This survey practice is done by the collection of 70 prescriptions as well as questionnaire on patients' age, gender, area, medication pattern, management option, awareness of people etc.

According to the table of gender of patients (Table no. 1) it can be seen that, male people (98.57%) are affected highly in this diseases than female. This is because genetically males are more susceptible than females.

This study reveals that this disease is highly dangerous to the age group people who are between 0-18 years of age and this can be expressed as percentage about 66.66% and the lower most affected group people are between 45-60 years of age and in percentage it is about 0%. (Table no. 2)

According to the data shown in (Figure no. 1), Dhaka is found most prevalent area among all districts and in percentage it is about 49%. On the other hand, least prevalent area is found as sylhet about 4% people.

The causes of hemophilia is found mainly genetically transferred gene carried from mother to fetus is about 89.83% for type A & 81.81% for type B. Other reason is found unknown. (Figure no. 2)

In Figure no. 3, the highest found symptoms among patients are knee joint swelling (34.28%). Some other symptoms also found in some cases like shoulder pain, blood in urine, bleeding gum, nosebleed patients are about (21.42%), (20%), (14.28%), (10%) accordingly.

Anti-hemophilic injection taking is found in greater content than freshly frozen plasma for both type A and type B patients about 79.66%, 72.72% and 20.33%, 27.27%. (Table no. 3)

The study also reveals the information about injection taking frequency of both type A and type B patients shown in figure no. 5 and is found daily frequency about 70.38% & 66.67%. Least common injection taking frequencies among patients are found monthly about 7.04% and 11.11%. (Figure no. 4)

According to the Table no. 4, it is shown that people acquaintance with this disease is very low; about (74.28%) have no knowledge on this bleeding disorder.

Mostly prescribed brand for the management of other complications is found Napadol (39%). There are also some other brands are found less common like Traxyl (Inj.) (26%), Ace (21%) & Traxyl (Cap.) (14%). (Figure no. 6)

It's a genetic disorder and it's a very rare diseases occurred mainly in male people. By doing some home diet we can also cure this diseases because prevention is better than cure. Making awareness among people is helpful to identify and manage this disease.

Many people in the world are suffering from bleeding disorders and they have not enough knowledge on hemophilia and related treatment options. Bangladesh is set to the joining the global community of hemophilia. Though the disease is incurable, but with proper knowledge and treatment it can be handled. The demand included blood fresh plasma and essential injections to the patients to cure this disorder. Here this study area is covered only limited hospitals, more study is needed to create awareness, observe nationwide condition as well as find out any other medication pattern for hemophilia patients.

Acknowledgments

Corresponding authors of this study article are thankful and grateful to the patients, patients' family, doctors, nurses, and hospital authority for their heartiest cooperation to conduct the study.

Reference

1. The hemophilia and von brand and platelet disease handbook.

2. Soucie JM, Evatt B, Jackson D, Hemophilia Surveillance System Project Investigators. Occurrence of hemophilia in the United States. American journal of hematology. 1998 Dec;59(4):288-94.

DOI:10.1002/(SICI)10968652(199812)59:4<288::AID-AJH4>3.0.CO;2

3. Genetics home reference; retrieved in 22 October, 2018.

4. www.medicalnewstoday.com; retrieved in 21 October, 2018.

5. Skinner MW. Haemophilia: provision of factors and novel therapies: World Federation of Hemophilia goals and achievements. British journal of haematology. 2011 Sep; 154(6):704-14.

doi:10.1111/j.1365-2141.2011.08765.x

6. Srivastava A, Brewer AK, Mauser-Bunschoten EP, Key NS, Kitchen S, Llinas A, Ludlam CA, Mahlangu JN, Mulder K, Poon MC, Street A. Guidelines for the management of hemophilia. Haemophilia. 2013 Jan;19(1):e1-47.

DOI: 10.1111/j.1365-2516.2012.02909.x

7. Jones P, Robillard L. The World Federation of Hemophilia: 40 years of improving haemophilia care worldwide. Haemophilia. 2003 Nov;9(6):663-9.

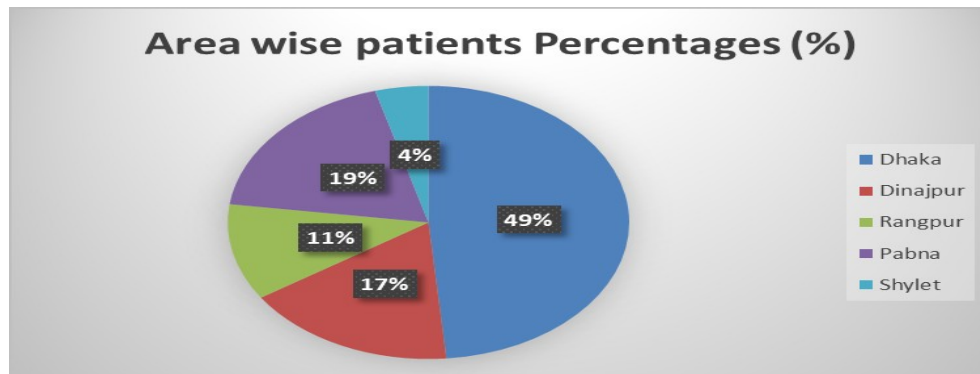
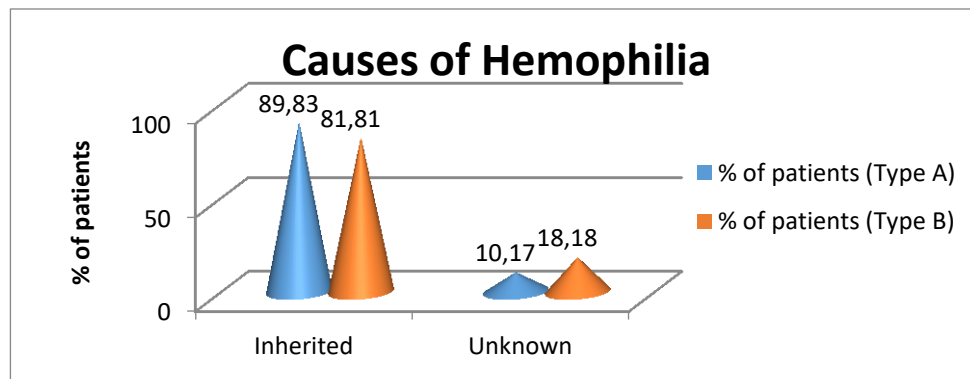
doi:10.1046/j.1351-8216.2003.00831.

Table 1. Prevalence of hemophilia disease according to gender

| Gender | Number of patients | Percentage (%) |
|--------|--------------------|----------------|
| Male | 69 | 98.57% |
| Female | 1 | 1.42% |
| Total | 70 | 100 |

Table 2.: Percentage of hemophilia patients (Type A & B) according to age.

| Age of patients | No. of patients of Type A | Percentages (%) | No. of patients of Type B | Percentages (%) |
|-----------------|---------------------------|-----------------|---------------------------|-----------------|
| 0-18 | 33 | 55.93% | 8 | 66.66% |
| 18-25 | 10 | 16.94% | 1 | 8.33% |
| 25-45 | 11 | 18.64% | 2 | 16.66% |
| 45-60 | 5 | 8.47% | 0 | 0% |
| Total | 59 | 100 | 11 | 100 |

Figure 1. Area wise distribution of patients.**Figure 2.** Causes of hemophilia (Type A & Type B)**Figure 3.** Comprehensive list of associated symptoms of Hemophilia patients (%)

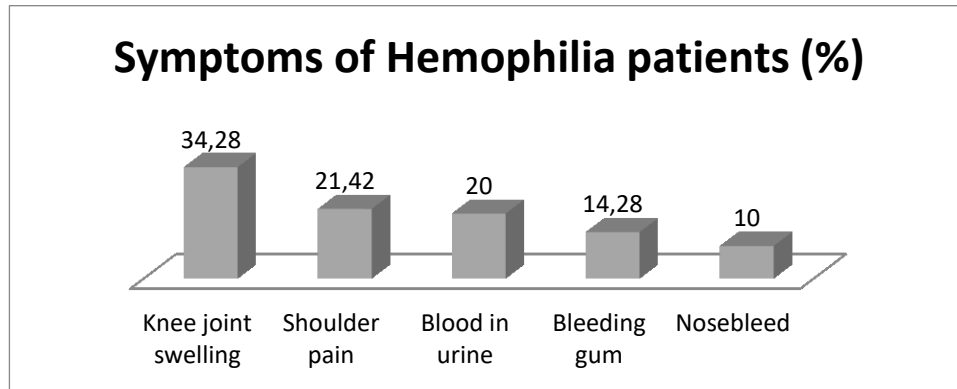


Table 3. Comprehensive list of medication pattern of patients (Type A & Type B)

| Medication type | Number of patients Type A | Percentages% | Number of patients Type B | Percentages% |
|----------------------------|------------------------------|--------------|------------------------------|--------------|
| Anti- hemophilic injection | 47 | 79.66 | 8 | 72.72 |
| Fresh frozen plasma | 12 | 20.33 | 3 | 27.27 |
| Total number | 59 | 100 | 11 | 100 |

Figure 4. Anti-hemophilic injection taking frequency of patients (%)

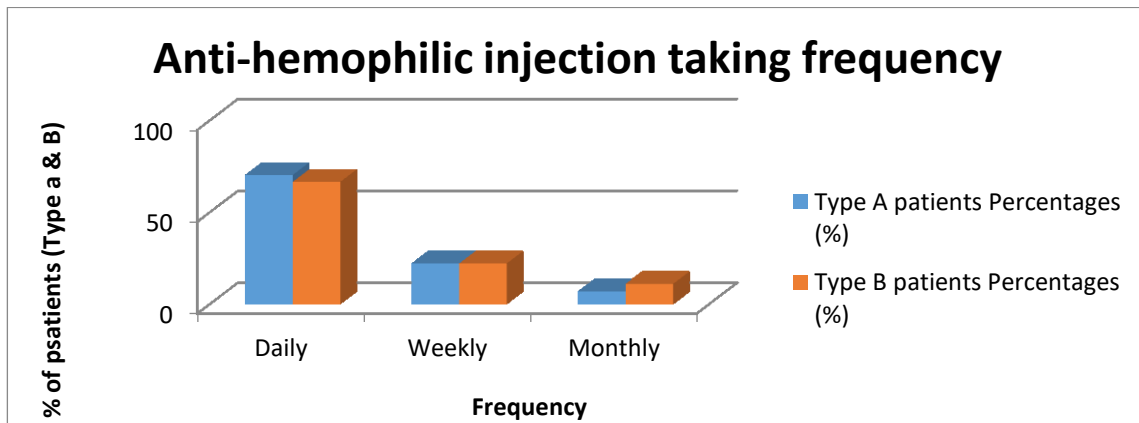


Figure 5. Fresh plasma taking frequency of patients (%)

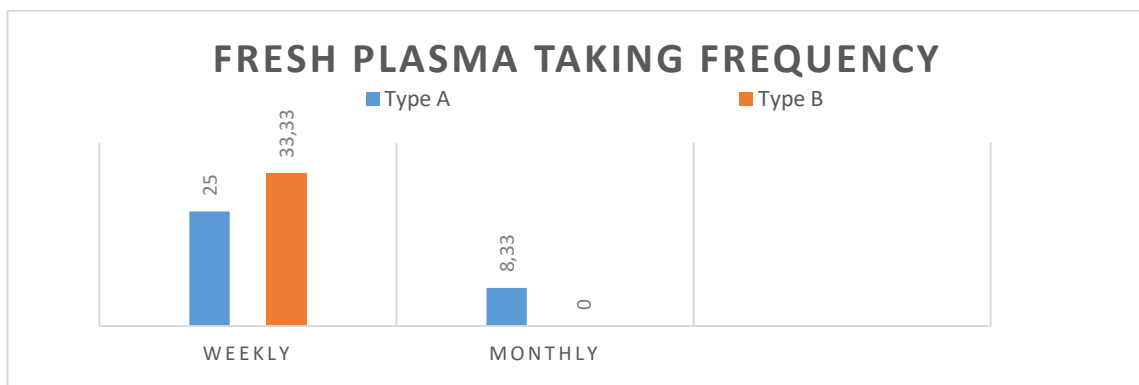


Table 4. Comprehensive list of Patients acquaintance (%)

| Patients acquaintance | Number of patients | Percentages (%) |
|-----------------------|--------------------|-----------------|
| Knowledge | 18 | 25.71 |
| No knowledge | 52 | 74.28 |
| Total Number | 70 | 100 |

Figure 6. Comprehensive list of another medications (%)