

## Estimation of Severity of Oral Health Problem(s) in Children with Different Types of Hemophilia by Socio-demographic Characteristics

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### ABSTRACT

**Introduction:** Hemophilia is an awfully distressing and life-long disorder and little is known about the influence of socioeconomic status on its severity. Life-long treatment of a person with hemophilia demands a sizable financial load. The mild form of hemophilia may frequently be left undiagnosed till their adolescent or even later in life, mainly if a patient did not have any exposure to surgery or extensive dental work or severe injury at an earlier age. Since hemophilia is a life-long disorder that creates great physical, emotional, economic and social problems for those afflicted [1]. Therefore, it is a real burning issue to introduce any suitable procedure to take care of them as efficiently as possible.

**Materials and Methods:** A hospital-based cross-sectional descriptive study was conducted at the Pedodontics Department of Faculty of Dentistry, Bangabandhu Sheikh Mujib Medical University (BSMMU), Bangladesh, from July 2015 to June 2016. The present study was performed to determine the oral manifestations and oral health behaviors of children with hemophilia according to the sociodemographic perspective. For this study, a purposive sampling method was applied. The hemophiliacs (either hemophilia A or B) children, (5-12 years old) came to the department of Pedodontics of BSMMU for seeking dental treatment, were selected as a study sample. A total of 54 hemophiliac cases were included in this study.

**Results:** The mean age of the study subject was 7.2 years. From the study result, it was marked that the blood group of hemophiliac patients was mostly 'O' positive (50%) whereas the least numbers of blood group were AB positive (5.5%). No one's blood group was (Rh) negative. 44.4% of study children had no detective family history of bleeding disorder whereas 55.6% had a strong family history. Only 13.3% respondent's mother was known carrier while 40% of families do not know about this. Amongst the parents, only 2 couples (3.7%) got married within relatives (first cousin). In terms of severity of the disease, study findings also described that the most number of study subjects were suffering from severe (44.4%) and moderate (38.9%) types of hemophilia. Except 1 no patient was capable of taking prophylaxis regularly. Among the participants, 47% belongs to the lower socio-economic status and the rest 53% from the lower middle class. No respondents were available from the upper middle as well as upper-class society.

**Conclusion:** From this study reflection it can be acclaimed to the concerned authority for providing factor VIII and IX at a cheaper rate or free for very poor patients. However, more study could be done focusing on the link of blood group as well as socio-economic status with hemophilia and its severity.

### Keywords

Oral health, Childrens, Hemophilia.

### Aims/Objectives

#### General Objective

To describe the oral manifestation and oral health behavior of hemophiliac children especially regarding socio-demographic

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perspective.

### Specific Objectives

- To find out the socio-demographic information of respondents with different stages of Hemophilia A and B.
- To find out the risk factors association among hemophiliac children for severity of oral manifestations.
- To find out the knowledge, attitude, and practice for among children respondents.
- To improve the provision of oral healthcare of an individual with hemophilia regardless of the socio-economic status.

### Introduction

Hemophilia is rare and the most common heritable genetic bleeding disorder [2]. This illness impairs the body's ability to make blood clots due to lack of certain protein responsible for blood clotting called clotting factor. As a consequence, easy bruising, lengthier bleeding after an injury takes place because of clotting factor deficiency. As in hemophiliac patient blood does not clot properly, it is life-threatening and an increased risk of bleeding inside the joints or the brain. Among 13 clotting factors, assist the blood to clot functioning with platelets, due to their deficiency cause the bleeding disorder. When Factor VIII or Factor IX or very rarely Factor XI is deficient in the bloodstream causes a bleeding disorder called hemophilia. According to the World Federation of Hemophilia (WFH), about 1 in 10,000 people (or 1 in 5,000 male births) for hemophilia A and 1 in 50,000 births for hemophilia B are born with this disease [3].

Hemophilia is caused by a defect/deficiency in the gene that determines the production of factor VIII or IX or XI. Since the factor VIII and IX are located on the X-chromosome they are also named as an Xlinked recessive disease [4]. In the royal families of Europe hemophilia was a common illness in the 1800s [5].

In children with hemophilia, bruising is very common though usually bruise is not caused for alarm, except when it is on the individual's head or neck. Apart from these some bleeds, such as bleeding in the head, throat, gut or iliopsoas, can be life-threatening and so requiring immediate treatment. The number of factors in the blood especially factor VIII or factor IX (on which the present study was concerned) states the severity of hemophilia. In this regard, hemophilia is categorized as mild, moderate, and severe according to the ranks of severity [6].

### Severe Hemophilia

When the amount of clotting factor in the plasma is of less than 1 percent then it falls into the severe category. Severe hemophiliacs usually bleed frequently, one to two times per week, into their muscles or joints [7]. They may often bleed spontaneously without any obvious reason. People with severe hemophilia and sometimes with moderate hemophilia, internal bleeding is common. Joint bleed is the most typical type of character of internal bleeding [8].

### Moderate Hemophilia

When clotting factor in the plasma remains between 1 and 5 percent

then it falls into the moderate category. In the moderate type of hemophilia, bleed less frequently that usually happens following an injury. However, this episode varies case to case and it might be possible for a moderate hemophiliac to bleed spontaneously.

### Mild Hemophilia

If clotting factor in the plasma remains between 5 and 40 percent then it indicates as mild hemophilia. In the case of mild hemophilia usually, bleeding occurs only as a result of surgery or major trauma [6].

At birth in mild to moderate hemophilia, except during undertaking circumcision, usually, no signs or symptoms have visualized. Later in life, the first symptoms are large bruises and hematomas occur following falls at the time of starting walking. Mild hemophiliac children may not have visible symptoms for several years. In contrast, in very mild hemophiliacs, heavy bleeding may happen even from a simple dental procedure, an accident, or surgery. The female carriers, using their one normal gene, able to produce enough clotting factors and thus can prevent serious bleeding problems, though some may not be capable of preventing bleeding.

There are several forms of bleeding disorder:

### Hemophilia A

The most common type of hemophilia is due to lack of Factor VIII. According to National Heart Lung and Blood Institute (NHLBI), 8 out of 10 people with hemophilia have hemophilia A. During pregnancy the level of factor VIII (not factor IX) has a tendency to rise.

### Hemophilia B

It is caused by the deficiency of Factor IX that is also termed as Christmas disease.

Hemophilia A and B are more common in males than females because of genetic transmission. As hemophilia A and B are both X-linked recessive disorders, females are rarely severely affected [9]. Few females with a nonfunctional gene on one of the X chromosomes may be mildly symptomatic [5]. There is spontaneous bleeding in both hemophilia A and B but a normal bleeding time, normal prothrombin time, normal thrombin time, but prolonged partial thromboplastin time.

### Hemophilia C

This is a very rare and mild type of bleeding disorder caused by lacking factor XI. Here, the patient does not experience spontaneous bleeding except after trauma or surgery. Hemophilia C is an autosomal inherited disease that affects males and females equally because this genetic defect is not related to the sex chromosome. Hemophilia C is commonly found in Ashkenazi Jews [5].

### Acquired Hemophilia

An extremely rare type of hemophilia may develop after birth when a new mutation occurs during early development or later in life. In this type of hemophilia, the patient's immune system

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forms antibodies against factor VIII or factor IX and thus these antibodies attack these factors. As a result, the body's immune system destroys its own naturally formed factor VIII and factor IX [9,10]. This disorder often settles with proper dealing. Acquired hemophilia might be connected with cancers, autoimmune disorders, and pregnancy [11,12].

If a person's immune system reacts to proteins present in factor(s) as harmful foreign elements as if the body has never seen them before, antibodies (also called inhibitors) are formed in the blood against this protein. When this ensues, antibodies start fighting against their own protein what they assume foreign factor protein. In this situation, it is very hard to control bleeding in the individual with hemophilia [6].

### **Para hemophilia**

Para hemophilia occurs due to not enough factor V production [13,14]. There are several other factor deficiencies including factor I, II, VII, X, XI, XIII and von Willebrand factor that also cause abnormal bleeding but the most severe forms of these types of deficiencies are even rarer in comparison to hemophilia A and B.

### **Diagnosis**

Hemophilia is diagnosed through a blood test from vein to measure its ability to clot and its levels of clotting factors [8].

### **Prevention**

Hemophilia is such a condition that is passed from a mother to her child and no way of knowing whether the baby has the condition before birth. Though prevention may occur by fertilizing an egg after removing it and then the embryo is tested before transporting to the uterus [8]. Again, now it might be possible to detect it (the fetal sex) at the gestational age of sixteenth week with a high degree of precision [15]. Hemophilia is not a curable disease but it can be possible to minimize symptoms and prevent future health complications by treating the patients in a proper way using prophylaxis.

Prophylaxis means before starting bleeding to prevent bleeding episodes usage of intravenous injection of clotting factor concentrates, made either from human blood or by recombinant methods or a combination of both on routinely [16]. The replaced clotting factors help to maintain an adequate level of factor VIII or IX in the bloodstream persistently when administered one injection of clotting factor two or three times a week. Hence, consequently, prophylaxis can be able to reduce or prevent joint destruction and increase the quality of life of hemophiliacs. Usually, it can be started between the ages of two and four years while the veins are well developed [17].

In case of mild hemophilia, clotting factors are not usually required while in moderate hemophilia clotting factors are essential only if bleeding occurs to prevent them [16]. But in severe hemophilia clotting factors are often recommended (two or three times a week) as a preventive measure and that may continue for life [16].

All of these are feasible after the development of blood products in the last few decades that can convert the view a lot especially in the well-off world. So regrettably, the choice of treatment fluctuates around the world. Cost affordability is a big deal in this respect. So, still, the cryoprecipitate is the main support of therapy in many underprivileged groups. Apart from this, many patients have a quiet chance to be infected with hepatitis and or HIV through the use of coagulation factor concentrates particularly when they receive donor's blood and before the introduction of physical methods of viral inactivation in the mid-1980s. In addition, coagulation factor concentrates are expensive however very much cost-benefit.

Bleeding in hemophilia mostly happens internally. The prevalent types of bleeding patterns include hemo-arthritis, muscle hematomas, epistaxis, gum bleeding, gastrointestinal bleeding, and hematuria. Among them, all the joints such as the knee, ankle, and elbow are most commonly affected. Recurrent hemo-arthritis and internal bleeding characterize the severe form and without timely and effective treatment the prognosis will be poor. Because of frequent bleeding into the joint without timely treatment the cartilage and the bone in the joint can be damaged, causing chronic arthritis and disability though now hemophilia even in severe form, maintaining almost normal life can be possible after proper treatment [6].

Because hemophilia is a life-long disorder which makes profound physical, emotional, economic and social problems for the afflicted individual, in the present study we have tried to perceive these areas especially the socioeconomic problem and the relationship with the severity of the disease [7,18]. To obtain the information we have assessed the effect of the disorder upon the individual by means of an interview as well as the severity of the disease in terms of socioeconomic cost.

### **Methods**

#### **Study Population**

The research participants were the children of 5-12 years old who were suffering from hemophilia A or B.

#### **Study Area**

Pedodontics Department of Faculty of Dentistry of BSMMU, Shahbagh, Dhaka-1000, Bangladesh.

#### **Study Design**

A cross-sectional descriptive and hospital-based study

#### **Sampling Technique**

For this study, a purposive sampling method was applied. The children who were known hemophiliacs (either A or B), came to the department of Pedodontics of BSMMU for seeking dental treatment, were selected as a study sample. No effort was made to exclude von Willebrand's disease in those with confirmed factors VIII (FVIII) deficiency.

We explained the parents/guardians of the children who met the study eligibility criteria about involving their children in the

study. Written consent and confidentiality were measured and emphasized during enrolment procedures.

### Inclusion Criteria

All individuals with mild to severe hemophilia (either A or B), between the age of 0-14 years, visited the study place, were eligible to participate.

### Exclusion Criteria

The children above the age of 14 years and not suffering from hemophilia were not included in this study.

**Sample Size:** The sample size was 54.

**Study Period:** 01-07-2015 to 30-06-2016: Total 12 months

### Data Collection Tools

Using a standardized, semi-structured questionnaire including demographic and clinical information, data was collected by the research assistant(s), recruited by the principal investigator. The co-investigator and the principal investigator trained up the research assistants in connection with patients' care to obtain accurate, reliable and valid data as well as protecting patient's confidentiality.

### Data Collection Procedure

Demographic and clinical details were collected from 54 children who agreed to participate in the study. It was a purposively sampled data, using maximum variation sampling (stratified by specific demographic location, ABO blood group, socioeconomic status, the severity of the disease and oral manifestation as well) in order to ensure a wide cross-section of the individual. As the study populations were too young to provide the information, all participants' guardians were invited to contribute valid information in a semi-structured questionnaire. In return, we provided them at least one free dental treatment as an enticement or gift.

Clinical examinations were conducted by the Principal Investigator and Co-investigator. The severity of hemophilia was categorized by the percentage of factors VIII and IX deficiency.

Data from each study participant was saved using the confidential and exclusive ID number. Confidentiality maintained through all data being stored securely and electronic data will be password-protected. Data was only be identified by an identification code, the key to which was stored individually and just accessible by the principal investigator.

### Statistical Method

The data from the questionnaire and the clinical examination was analyzed using SPSS version 21. Descriptive statistics (Frequency, percentage, mean) was reported. Descriptive statistics included socio-demographic characteristic, ABO blood group, clinical characteristics (mild to severity) and health status of the patients were used to summarize the data. Responses were grouped

according to the severity of the disease, ABO blood grouping, affordability for the treatment, socio-economic distribution. The association between the oral health status of hemophiliac and the socio-demographic characteristic and other study variables were considered.

### Results Obtained

A total of 54 hemophiliac children aged 5-12 years old, with mean age 7 years, were enrolled as a sample for this study involving their parents. Among them, 88.9% (48) were from male sex whereas 11.1% were from female sex. Of all study subjects, 88.9% had Hemophilia A except 11.1% who had hemophilia B.

Demographic data		%
Sex	Male	88.9
	Female	11.1
Age (Years)		Mean
5 to 12		7.2
Body wt. (Kg)		Mean
11 to 37		18.5
Clinical data	Type of Hemophilia	%
	A	88.9
	B	11.1
Hemophilia severity	Mild	16.7
	Moderate	38.9
	Severe	44.4

Table 1 lists the demographic and clinical data of patients with hemophilia A and B.

According to Figure 1, the distribution of blood group among the study participants was 22.2% of respondents had 'A' positive blood group, 22.2% had 'B' positive, 5.5% had 'AB' positive and 50% had 'O' positive blood group.

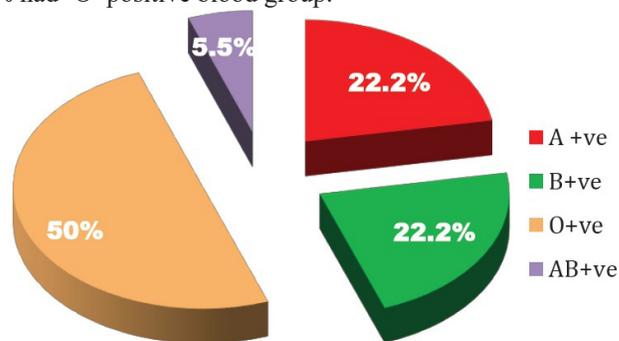
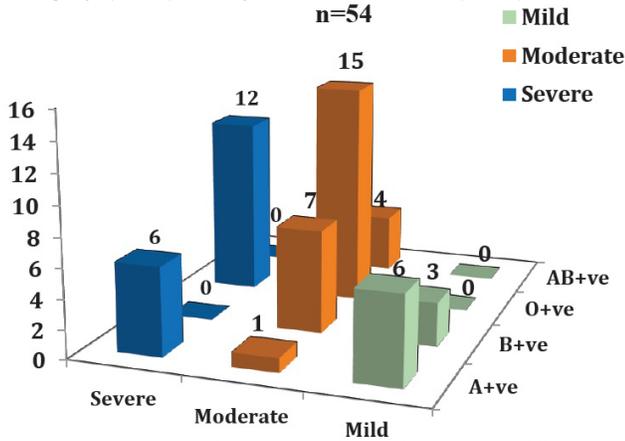


Figure 1: Percentage of Blood Group Among Hemophiliac Child.

Figure 2 showing the distribution of the blood group in different stages of hemophilia. From the study result, it was also evident that the frequency of 'O' positive blood group constituted the maximum (almost) both in moderate and severe cases of hemophiliac patients which were absent in mild cases.

In most children (53.3%) hemophilia were first detected between the age of 11 months to 3 and half years of age, 46.6% of the

participant's first sign was purpuric or bruising spot throughout the body, while 20% had gum bleeding following trauma, 20% had pain and swelling of joint(s) and only a few cases were detected due to the incidence of epistaxis (6.6%), non-stop bleeding after minor injury (6.6%) and by routine blood test (6.6%).

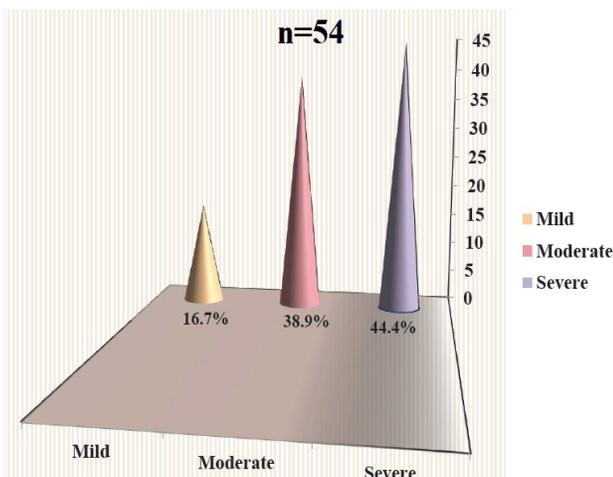


**Figure 2:** Showing the Distribution of Blood Group in Different Stages of Hemophilia.

From the study result, it was evident that the birth order of the most study children (44.4%) were 1 while 38.9% were 2 and 16.7% were 3.

44.4% of study children had no detective family history of bleeding disorder whereas 55.6% had a strong family history. Only 13.3% respondent's mother was known carrier while 40% of families do not know about this. Amongst the parents, only 2 couples (3.7%) got married within relatives (first cousin). 86.6% of the siblings of the victims do not have this problem but 13.3% siblings were also the sufferer for the same problem.

According to Figure 3, the study data explained the larger number of study children 24 (44.4%) were in a severe category whereas 21 (38.9%) were in a moderate group and 9 (16.7%) were the mild type.



**Figure 3:** Percentage of different categories of hemophilia in terms of severity.

Although the therapy of hemophilia is now allowing many severe

hemophiliacs to achieve maturity in this study except 1 child nobody received any prophylaxis regularly, 53.3% received very irregularly and 46.6% received only during their need. Among the study children, only 26.6% were able to afford the factor when they need but the rest 73.4% cannot afford it and so received FFS even if they need the factor very urgently.

Figure 4 viewing 47% belong to the lower socio-economic status whereas the rest 53% were from the lower middle class. No respondents were available from the upper middle as well as upper-class society.

Affordability to take Factor when they need	Percent (%)
Yes	26.6%
No	73.4%
<b>Total</b>	<b>100%</b>

n=54

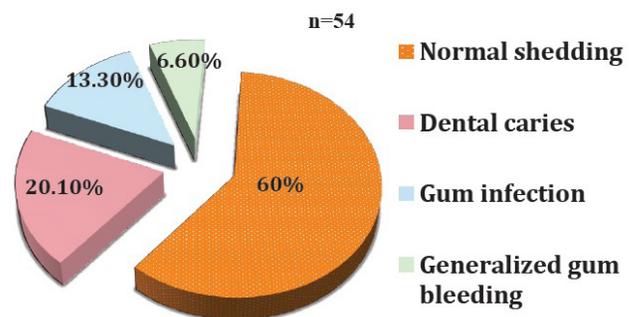
### Socio Economic Status



**Figure 4:** Displaying the distribution of socioeconomic class among the study population.

Figure 5 presenting 60% of the study children arrived at the Department of Pedodontics for seeking various oral and dental cares, such as gum bleeding due to mild trauma by the loose deciduous tooth at its normal shedding time, 20.1% came for uninfected dental caries, 13.3% came with gum infection due to dental caries and only 6.6% came for generalized gum bleeding.

### Complaints of Patients



**Figure 5:** Showing the distribution of different dental complaints of the study subjects.

### Discussion

Increasing evidence directs the association between ABO blood group determinants and hemostasis. In this study, the investigator observed a relationship between ABO blood groups and factor

VIII and IX among hemophiliacs. According to the study findings, 22.2% of respondents had 'A' positive blood group, 22.2% had 'B' positive, 5.5% had 'AB' positive and 50% had 'O' positive blood group. So, the number of 'O' blood group is significantly more than any other blood groups. Other studies have consistently reported that subjects of 'O' blood group have lower plasma von Willebrand factor (VWF) levels than non-O individuals [19]. Many investigations have proved the relationship between ABO blood groups with the hemophiliac cohort. Although Torghabeh HM and et al. found no relation between ABO blood groups and factor VIII and IX inhibitors [20]. Several studies have recognized the impact of ABO blood groups on plasma VWF levels [17,21,22]. Although a number of studies have established the effect of ABO blood group on plasma levels of von Willebrand factor (VWF), it is still basically unknown in terms of the nature of this association and its clinical significance and few studies showed the association between ABO blood group with the hemophilia A and B.

The effect of hemophilia including its treatment has not yet been studied well focusing on social status. After the availability of the safe, effective and purified coagulation factor (FVIII & FIX) concentrates that could be used as prophylaxis [23], maximum research advocates that amongst children, adolescents, and adults with hemophilia, social status, and well-being is not compromised rather than comparable to that of the general population [15]. Though the main objective of the present study was in the quest for the association between the severity of the disease and the socio-economic status the present study sample did not contain any participant from the high-socio-economic group. So, the present study findings emphasize the necessity of conducting an additional study with the precise direction in terms of using power sample as well as performing a longer prospective study.

## Conclusion

From the present study result, a clue was evident about the association between hemophiliac and blood group. In this study, the study children who had 'O' positive blood group constituted the highest number in terms of severity as well as among all study participants. Since the sample size was not big enough because it was a hospital-based study and so the obtained data may not represent the whole country. Therefore, a larger further longitudinal research is suggested to settle the conclusion of the present study by exploring all relevant variables deeper into its mechanisms.

## Recommendation

As hemophilia is not a curable disease that requires life-long care, produces a bulk of the burden for a family. Because of the high cost it is not possible for most of the patients to maintain their condition using prophylaxis; especially it is very true in the context of Bangladesh. In this regard, the government can focus on providing free factors (VIII or IX) or supply them at a cheaper rate particularly for the underprivileged population.

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