

Original article:

Study of anemia among hemophilia patients

Rafiqul H¹, Biswas DA², Quraishi SA³

Abstract

Background: Almost all hemophilia patients are treated with fresh frozen plasma and other alternatives regardless of anemic status, so it is important to identify the patients who are chronically anemic and thereby those cases to be treated accordingly. As no previous work and research regarding study of anemia among hemophilia patients were done in our country and abroad as far as I know, so data regarding this study from online source could not be compiled here. **Objective:** The objective of the study was to evaluate the degree of anemic status among hemophilia patients who were receiving FFP for long time. **Materials and Method:** It was a prospective type of observational study. The study was carried out in the Department of Transfusion Medicine, BSMMU, Dhaka, Bangladesh. This study was conducted from July 2011 to June 2012 for a period of one (1) year. 60 patients with hemophilia were selected from patients attending at day care unit of Transfusion Medicine dept. of BSMMU. **Results:** Among 60 patients, according to degree of anemia majority cases are moderately anemic which were 35(58.33%) cases followed by mildly anemic which were 15(25.00%) cases, Severely anemic were 2(3.33%). Non anemic cases were 10(16.67%).

Bangladesh Journal of Medical Science Vol. 17 No. 04 October'18. Page : 573-575

DOI: <http://dx.doi.org/10.3329/bjms.v17i4.38318>

Introduction:

Hemophilia is a coagulation disorder that delays the blood clotting process. People with this condition experience prolonged bleeding or oozing following an injury, surgery, or having a tooth pulled. In severe cases of hemophilia, heavy bleeding occurs after minor trauma or even in the absence of injury (spontaneous bleeding). Serious complications can result from bleeding into the joints, muscles, brain, or other internal organs. Milder forms of hemophilia do not involve spontaneous bleeding, and the condition may not become apparent until abnormal bleeding occurs following surgery or a serious injury.

The conventional treatment of haemophilia^{1,2} is Fresh Frozen Plasma. FFP is defined as the fluid portion of one unit of human blood that has been centrifuged, separated from PRP, and frozen solid at -18° C (or

colder) within 6 hours of collection. Other single-donor plasma units, either frozen or liquid, may be substituted for FFP.

Although the recombinant factor VIII and IX^{3,4,5,6} is available but it is not affordable due to higher cost. In our country most patients of hemophiliacs are treated with FFP. FFP needed in large volume and transfusion of large volume of FFP in anemia is a danger. The study is carried out with an intention to assess Hb concentration in hemophiliac patients who are chronically receiving FFP due to multiple haemorrhage and to evaluate the adverse reaction to minimize subsequent complication and thereby helping physician to rational management of haemophilia with best possible safety for the people of our country.

1. Rafiqul Haque, Assistant Professor, Department of Transfusion Medicine, Ibn Sina Medical College. Email: rafiqulhaquefiroz@yahoo.com
2. Dr. Dannish Arefin Biswas, Associate Professor, Department of Transfusion Medicine, Sir Salimullah Medical College, Dhaka, email: dr.dannish.arefin@gmail.com
3. Dr. Shafiul Alam Quraishi, Assistant Professor, Department of Pediatrics, Ibn Sina Medical College

Correspondence to: Rafiqul Haque, Assistant Professor, Department of Transfusion Medicine, Ibn Sina Medical College. Email: rafiqulhaquefiroz@yahoo.com

Materials and Method: It was a prospective type of observational study. The study was carried out in the Department of Transfusion Medicine, BSMMU, and Dhaka, Bangladesh. This study was conducted from July 2011 to June 2012 for a period of one (1) year. : Patients who were diagnosed as cases of hemophilia attending in the day care service of the department of Transfusion Medicine, BSMMU for management. Hb% estimated by colorimeter^{7,8,9}. 60 patients with hemophilia will be selected from patients attending at day care unit of Transfusion Medicine dept. of BSMMU.

Ethical Clearance: This study was approved by Institutional Review Board (IRB) of Bangabandhu Sheikh Mujib Medical University.

Results:

A total number of 60 patients who were diagnosed as a cases of Hemophilia at any age of both sexes and who were admitted in BSMMU at day care center of Transfusion Medicine department were enrolled in this study.

Table 1 : Distribution of the study population according to degree of anemia (n=60)

General appearance	Frequency	Hb%	Percent
Mildly anemic	15	< 11 gm/dl	25.00
Moderately anemic	35	6-10 gm/dl	58.33
Severely anemic	02	< 6 gm/dl	3.33
Non anemic	08	> 12.5 gm/dl	13.33
Total	60		100.0

Table1: shows the distribution of study population according to anemic status. Among the 60 patients maximum cases are moderately anemic which is 35(58.33%) cases followed by mildly anemic which is 15(25.00%) cases, Severely anemic is 2(3.33%). Non anemic cases are 10(16.67%).

Table 2 : Distribution of the study population according to number of hemorrhagic sites. (n=60)

N u m b e r hemorrhagic site	Frequency	Percentage
Single	20	33.33
Multiple	40	66.67
Total	60	100

Table II : shows the distribution of study population according to number of hemorrhagic sites. Among 60 cases 20 (33.33%) cases have hemorrhage at single joint and 40 (66.67%) cases have hemorrhage at more than one sites.

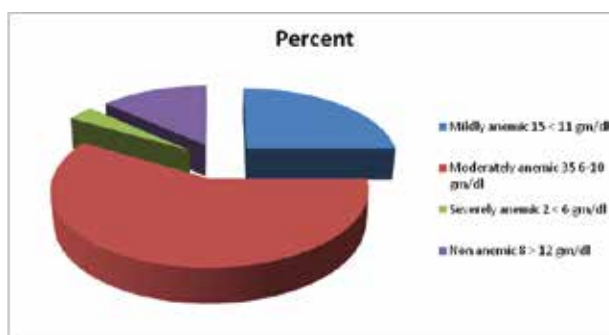


Fig 1: Pie chart of study population according to anemic status

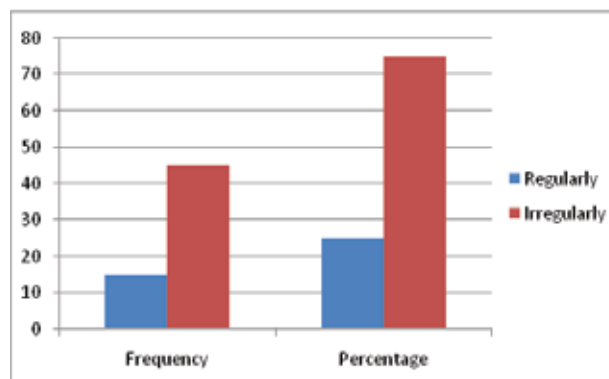


Fig 2 : Bar diagram of study population according to checking Hb% status

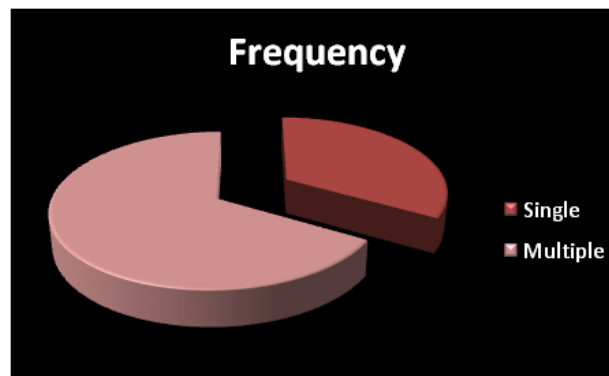


Fig 3 : pie chart of study population according to number of hemorrhagic sites

Discussion:

Hemophilia ^{10,11,12} is a genetic disease linked to a defective gene on the X chromosome. Chromosomes come in pairs -- women have two X chromosomes while men have one X and one Y chromosome. Patient with hemophilia are frequently treated with FFP but it has been seen most patients frequently present with bleeding in joints and mucus membrane have mild to severe anemia. but practically they are not treated for the correction of anemia. As a result we found that hemophiliac patients are usually chronically anemic.

The distribution of study population according to anemic status (Hb%) which was measured by

colorimeter was recorded in this study. Among 60 patients maximum cases were moderately anemic which is 35(58.33 %) cases followed by mildly anemic and severely anemic which are 15(25 %) and 02(3.33 %) cases respectively. Non anemic cases were 08(13.33 %). This finding suggests that majority of the patients were chronically anemic who are not corrected by appropriate therapy. The distribution of study population according to checking Hb% regularly or irregularly was also recorded. Among 60 patients majority 45(75 %) cases did not check Hb% regularly. Only 15(25 %) cases used to check Hb% regularly. This finding reveals that in majority cases patient's anemic status were neither evaluated nor corrected

Limitation of the study:

There were limitations in the present study. The major difficulty encountered during this study was missing of the patients for proper follow up. Majority of the patients left the day care center immediately after taking transfusion. There was another limitation faced that estimation of Hb% was performed by colorimetric method not by automated hematology cell analyzer.

Recommendation:

Further study is recommended with following proposal :

- This study will help the physicians and clinicians to measure Hb% before and after transfusion of FFP and thereby guide the clinician whether correction of anemia is needed during session of FFP transfusion.
- A large scale multi centered study should be done.
- Study period should be prolonged.
- As there is an increasing demand for FFP for transfusion in hemophiliac patients hospital in district level should establish a instrumental setup to prepare FFP and transfuse there, so thereby reduce intense work load in few center in Dhaka like BSMMU.

Conclusion:

In this study it was found that majority of the hemophilia patients receiving FFP were moderately anemic. The study suggest that hemophilia patients need to be investigated for anemia along with the FFP therapy. This study will help the physicians and clinicians to measure Hb% before and after transfusion of FFP and thereby guide the clinician whether correction of anemia is needed during session of FFP transfusion.

Conflict of interest : None

Reference:

- Bolton-Maggs PH, Pasi KJ. Haemophilias A and B. *Lancet*. 2003 May 24;**361**(9371):1801-9. Review. PubMed citation
- Franchini M. Acquired hemophilia A. *Hematology*. 2006 Apr;**11**(2):119-25. Review. PubMed citation
- Giangrande P. Haemophilia B: Christmas disease. Expert Opin Pharmacother. 2005 Aug;**6**(9):1517-24. Review. PubMed citation
- Graw J, Brackmann HH, Oldenburg J, Schneppenheim R, Spannagl M, Schwaab R. Haemophilia A: from mutation analysis to new therapies. *Nat Rev Genet*. 2005 Jun;**6**(6):488-501. Review. PubMed citation
- Oldenburg J, El-Maarri O. New insight into the molecular basis of hemophilia A. *Int J Hematol*. 2006 Feb;**83**(2):96-102. Review. PubMed citation
1. Plug I, Mauser-Bunschoten EP, Bröcker-Vriends AH, van Amstel HK, van der Bom JG, van Diemen-Homan JE, Willemsse J, Rosendaal FR. Bleeding in carriers of hemophilia. *Blood*. 2006 Jul 1;**108**(1):52-6. Epub 2006 Mar 21. PubMed citation
 2. Drabkin D.L et - *j. Biol Chem*.1932 ; **98** :719
 3. International Committee for Standardisation in Haematology – *Brit j. Haemat*. 1967; **13**: 71
 4. ZIJLSTRAN N.C.- *clin. cgim Acta* 1960; **5**: 719
Gene Review: Hemophilia A
Gene Review: Hemophilia B
- 12). I. Wallis JP, Dzik S. Is fresh frozen plasma overtransfused in the United States? Transfusion* 2004;**44**:1674-5.