International Journal of Medical Research Professionals

*Original Article*

# Study of Retrospective Analysis of Patients with Bleeding Disorders Visited in a Tertiary Care Hospital

**Pallavi Mishra[1], Ashutosh Mishra[2*]**

[1]Assistant Professor, Department of Medicine,
Major SD Singh Medical College, Farrukhabad, UP, India.

[2*]Ex-Associate Professor, Department of Medicine,
Gian Sagar Medical College, Patiala, Punjab, India.

**Article History**
Received: 04 July 2015
Revised: 19 July 2015
Accepted: 10 Aug 2015

***Correspondence to:**
Dr. Ashutosh Mishra,
Ex-Associate Professor,
Department of Medicine,
Gian Sagar Medical College, Patiala,
Punjab, India.

## ABSTRACT

**Background:** Inherited bleeding disorders lead to a lifelong bleeding tendency. The lack of sufficient treatment product for pain relief, for life threatening hemorrhagic episodes, and for the prevention of disability results in orthopedic morbidity, premature mortality, and extensive out-of-pocket expenditure. The present study was conducted to retrospectively evaluate patients with bleeding disorders.

**Materials and Methods:** In present study; a total of 300 patients were evaluated retrospectively. Before the commencement of the stud ethical approval was taken from the Ethical Committee of the institute. All the patients registered were included in the study. The data was collected using a questionnaire including age, gender, diagnosis, severity of disorder. The severity of the disease was based on plasma factor levels and the clinical manifestations of the disease; and divided into severe (less than one per cent of normal activity), moderate (one to five per cent of normal activity) and mild (6-30% of normal activity).

**Results:** A total of 300 patients had registered with inherited bleeding disorders in hospital. Inherited bleeding disorders identified in these patients included hemophilia A, hemophilia B, vWD, factor VII deficiency, factor V deficiency, factor X deficiency, dysfibrinogenemia, afibrinogenemia, factor XIII deficiency: and platelet function defects. Maximum prevalence was of haemophilia A (47.66%) followed by vWD (18.66%). Regarding disease severity, 62 patients (20.66%) had severe disease, 135 (45%) had moderate disease and mild disease was seen in 103 patients (34.33%).

**Conclusion:** The present study concluded that maximum prevalence was of haemophilia A followed by vWD. Regarding disease severity, 20.66% had severe disease, 45% had moderate disease and mild disease was seen in 34.33%. Maximum mild cases were of vWD followed by Hemophilia A. Maximum moderate cases were of Hemophilia A followed by Glanzmann's thrombasthenia. Maximum severe cases were of Hemophilia A followed by Hemophilia B.

**KEYWORDS:** Bleeding Disorders, Haemophilia A, vWD, Glanzmann's Thrombasthenia, Hemophilia B.

## INTRODUCTION

Hemostasis is the process by which bleeding is arrested after injury to blood vessels. It is a delicate multiphase process that involves interactions between the blood vessels, platelets, and coagulation factors. A defect in any of these phases of coagulation can result in a bleeding problem which may be inherited or acquired.[1-4]

Bleeding disorders reportedly affect 1 in 1000 individuals globally.[5] The most prevalent bleeding disorders are hemophilia A and B[6-8] and von Willebrand disease.[9-10] Hemophilia A and B are coagulation disorders caused by a deficiency of clotting factors VIII and IX, respectively. The coagulation factor level in the

body and the genotype of the patient determine the frequency of bleeding.[7,11,12] Hemophilia A affects 1 : 5000–10,000 males, while hemophilia B affects 1 : 50,000–100,000 males.[13] von Willebrand disease (VWD) is another bleeding disorder, which is an inherited disorder that is caused by deficiency or dysfunction of VWF. The prevalence of VWD has been estimated in several countries on the basis of the number of symptomatic patients seen at hemostasis centers and ranges from about 23 to 110 per million population (0.0023–0.01%).[14] Moreover, most mild bleeding disorders are often unrecognized, as patients bleed only during stress periods or with surgery and medical procedures.[15,16] The treatment of hemophilia is based on clotting factor replacement therapy. The most used treatments for hemophilia are "on-demand" therapy and factor prophylaxis. During a bleeding episode, clotting factor can be given "on-demand" to control and stop the bleeding. During prophylactic treatment, patients receive regular factor concentrate replacement therapy, which can lead to better clinical outcomes, with a lower overall impact on quality of life; however, this type of treatment is more expensive than "on-demand" therapy.[17,18] The present study was conducted to retrospectively evaluate patients with bleeding disorders visited in a hospital.

## MATERIALS AND METHODS

In the present study, a total of 300 patients were evaluated retrospectively. Before the commencement of the study ethical approval was taken from the Ethical Committee of the institute. All the patients registered were included in the study. The data was collected using a questionnaire including age, gender, diagnosis, severity of disorder. The severity of the disease was based on plasma factor levels and the clinical manifestations of the disease; and divided into severe (less than one per cent of normal activity), moderate (one to five per cent of normal activity) and mild (6-30% of normal activity).

**Table 1: Distribution of patients according to bleeding disorders**

| Bleeding disorder | N(%) |
|---|---|
| **Hemophilia A** | 143(47.66%) |
| **Hemophilia B** | 32(10.66%) |
| **vWD** | 56(18.66%) |
| **Factor VII deficiency** | 2(0.66%) |
| **Factor V deficiency** | 1(0.33%) |
| **Factor X deficiency** | 1(0.33%) |
| **Factor VIII deficiency** | 7(2.33%) |
| **Dysfibrinogenemia** | 1(0.33%) |
| **Afibriongenemia** | 12(4%) |
| **Glanzmann's thrombasthenia** | 33(11%) |
| **Bernard Soulier syndrome** | 4(1.33%) |
| **Undiagnosed** | 8(2.66%) |
| **Total** | 300(100%) |

**Table 2: Disease Severity in Bleeding Disorders**

| Bleeding disorder | Severity | | | |
|---|---|---|---|---|
| | **Mild** | **Moderate** | **Severe** | **Total** |
| **Hemophilia A** | 28(27.18%) | 63(46.66%) | 52(83.87%) | 143(47.66%) |
| **Hemophilia B** | 11(10.67%) | 16(11.85%) | 5(8.06%) | 32(10.66%) |
| **vWD** | 34(33%) | 21(15.55%) | 1(1.61%) | 56(18.66%) |
| **Factor VII deficiency** | 0(0%) | 0(0%) | 2(3.22%) | 2(0.66%) |
| **Factor V deficiency** | 1(0.97%) | 0(0%) | 0(0%) | 1(0.33%) |
| **Factor X deficiency** | 1(0.97%) | 0(0%) | 0(0%) | 1(0.33%) |
| **Factor VIII deficiency** | 3(2.91%) | 4(2.96%) | 0(0%) | 7(2.33%) |
| **Dysfibrinogenemia** | 0(0%) | 1(0.74%) | 0(0%) | 1(0.33%) |
| **Afibriongenemia** | 5(4.85%) | 5(3.70%) | 2(3.22%) | 12(4%) |
| **Glanzmann's thrombasthenia** | 11(10.67%) | 22(16.29%) | 0(0%) | 33(11%) |
| **Bernard Soulier syndrome** | 1(0.97%) | 3(2.22%) | 0(0%) | 4(1.33%) |
| **Others** | 8(7.7%) | 0(0%) | 0(0%) | 8(2.66%) |
| **Total** | 103(34.33%) | 135(45%) | 62(20.66%) | 300(100%) |

## RESULTS

A total of 300 patients had registered with inherited bleeding disorders in hospital. Inherited bleeding disorders identified in these patients included hemophilia A, hemophilia B, vWD, factor VII deficiency, factor V deficiency, factor X deficiency, dysfibrinogenemia, afibrinogenemia, factor XIII deficiency: and platelet function defects.

Maximum prevalence was of haemophilia A (47.66%) followed by vWD (18.66%). Regarding disease severity, 62 patients (20.66%) had severe disease, 135 (45%) had moderate disease and mild disease was seen in 103 patients (34.33%). Maximum mild cases were of vWD (33%) followed by Hemophilia A (27.18%). Maximum moderate cases were of Hemophilia A (46.66%) followed by Glanzmann's thrombasthenia (16.29%). Maximum severe cases were of Hemophilia A (83.87%) followed by Hemophilia B (8.06%).

## DISCUSSION

World federation of hemophilia surveyed 89% of world population in 2007, and reported 2,13,904 identified individuals with hereditary blood coagulation disorders (HBCDs) that included hemophilia A and B, Von Willebrand disease (vWD), and other bleeding disorders such as hereditary thrombotic tendencies, anomalies in fibrinogen to fibrin conversion, contact factor, extrinsic, and common pathway factor deficiencies.[19]

A total of 300 patients had registered with inherited bleeding disorders in hospital. Inherited bleeding disorders identified in these patients included hemophilia A, hemophilia B, vWD, factor VII deficiency, factor V deficiency, factor X deficiency, dysfibrinogenemia, afibrinogenemia, factor XIII deficiency: and platelet function defects. Maximum prevalence was of haemophilia A (47.66%) followed by vWD(18.66%). Regarding disease severity, 62 patients (20.66%) had severe disease, 135 (45%) had moderate disease and mild disease was seen in 103 patients (34.33%). Maximum mild cases were of vWD (33%) followed by Hemophilia A (27.18%). Maximum moderate cases were of Hemophilia A (46.66%) followed by Glanzmann's thrombasthenia (16.29%). Maximum severe cases were of Hemophilia A (83.87%) followed by Hemophilia B (8.06%).

El-Bostany et al. assessed the local prevalence of some inherited bleeding disorders in pediatric patients which involved 43 children with various bleeding manifestations recruited from a children's hospital in Cairo, Egypt, and Jeddah, Kingdom of Saudi Arabia. Of these, 12 (27.9%) had VWD, 11 (25.5%) had hemophilia A, three (7%) had hemophilia B, seven (16.3%) had platelet disorders, and 10 (23.3%) had bleeding of undiagnosed cause.[20] Ahmed et al. reported 34 cases of inherited bleeding disorders from Eastern Province of Saudi Arabia; of these, 15 had hemophilia, one had factor VII deficiency, one had factor X deficiency, 12 had Glanzmann thrombasthenia, and five had unidentified platelet function disorders.[21]

The common inherited bleeding disorders identified include hemophilia A, hemophilia B and vWD; majority of the patients have mild to moderately severe disease in the studies conducted by Zhang L et al (2003) and Manisha M et al (2002).[22,23]

Sajid et al. reported 37.2% (79) mild, 41% (87) moderate, 21.6% (46) severe Haemophilia A out of 212 patients of Haemophilia A.[24]

In a study by Ahmed et al. 77.8% cases of severe hemophilia A. 14.4% of moderate hemophilia A, and 7.75% cases of mild hemophilia A were reported and hemophilia B patients have 69.6% severe, 19.2% moderate and 11.2% mild disease.[25]

## CONCLUSION

The present study concluded that maximum prevalence was of haemophilia A followed by vWD. Regarding disease severity, 20.66% had severe disease, 45% had moderate disease and mild disease was seen in 34.33%. Maximum mild cases were of vWD followed by Hemophilia A. Maximum moderate cases were of Hemophilia A followed by Glanzmann's thrombasthenia. Maximum severe cases were of Hemophilia A followed by Hemophilia B.

## REFERENCES

1. Handin RI, Lux SE, Stossel TP. Blood Principles and Practice of Hematology. Philadelphia: J.B. Lippincott Company; 1995. pp. 9–72.

2. Ogedegbe HO. An Overview of Hemostasis. Lab Med. 2002;12(33):948–53.

3. Bick RL. Disorders of Thrombosis and Hemostasis Clinical and Laboratory Practice. 3rd edition. USA: Lippincott William's and Wilkins; 2002. Clinical assessment of patients with Hemorrhage; 1–37.

4. Schiffman F.J. Hematologic Pathophysiology. Lippincottts Pathophysiology series. Philadelphia and New York: Lippincott-Raven Publishers; 1998. Hemostasis and Thrombosis; 161–223.

5. Skinner MW. WFH: closing the global gap—achieving optimal care. Haemophilia 2012;18(Suppl. 4):1–12.

6. Lee CA. Historical introduction. In: Berntorp EE, Lee CA, Hoots WK,eds. Textbook of Hemophilia. (2nd ed.). West Sussex, UK: Wiley-Blackwell, 2010.

7. van den Berg HM, Fischer K. Phenotypic-genotypic relationship. In: Berntorp EE, Lee CA, Hoots WK, eds. Textbook of Hemophilia. (2nd ed.). West Sussex, UK: Wiley-Blackwell, 2010.

8. Mannucci PM, Tuddenham EG. The hemophilias—from royal genes to gene therapy.N Engl J Med2001;344:1773–9.

9. Rodeghiero F, Castaman G. von Willebrand disease: epidemiology. In: Berntorp EE, Lee CA, Hoots WK,eds. Textbook of Hemophilia. (2nd ed.). West Sussex, UK: Wiley-Blackwell, 2010.

10. Srivastava A, Rodeghiero F. Epidemiology of von Willebrand disease in developing countries. Semin Thromb Hemost 2005;31:569–76.
11. Berntorp E, Shapiro AD. Modern haemophilia care. Lancet 2012;379:1447–56.
12. Bolton-Maggs PH, Pasi KJ. Haemophilias A and B. Lancet 2003;361:1801–9.
13. W. F. O. Hemophilia. World federation of hemophilia report on the annual global survey 2006, in World Federation of Hemophilia 1425 René Lévesque Boulevard West, Suite 1010, Montreal, Canada.
14. W. L. Nichols, M. E. Rick, T. L. Ortel et al. Clinical and laboratory diagnosis of von Willebrand disease: a synopsis of the 2008 NHLBI/NIH guidelines. American Journal of Hematology 2009; 84(6): 366–70.
15. C. Biron-Andréani, B. Mahieu, A. Rochette et al. Preoperative screening for von Willebrand disease type 1: low yield and limited ability to predict bleeding. Journal of Laboratory and Clinical Medicine 1999; 134(6): 605–9.
16. M. Bowman, W. M. Hopman, D. Rapson, D. Lillicrap, M. Silva, and P. James. A prospective evaluation of the prevalence of symptomatic von Willebrand Disease (VWD) in a pediatric primary care population. Pediatric Blood & Cancer 2010; 55(1): 171–3.
17. Paisley S, Wight J, Currie E, Knight C. The management of inhibitors in haemophilia A: introduction and systematic review of current practice. Haemophilia. 2003;9:405–17.
18. Elander J. A review of evidence about behavioural and psychological aspects of chronic joint pain among people with haemophilia. Haemophilia. 2014;20:168–75.
19. Adeyemo TA, Adeyemo WL, Adediran A, Akinbami AJ, AkanmuAS. Orofacial manifestations of hematological disorders: Anemia and hemostatic disorders. Indian J Dent Res 2011;22:454-61
20. E. A. El-Bostany, N. Omer, E. E. Salama, E. A. El-Ghoroury, and S. K. Al-Jaouni. The spectrum of inherited bleeding disorders in pediatrics. Blood Coagulation & Fibrinolysis 2008; 19(8): 771–5.
21. M. A. M. Ahmed, M. O. Al-Sohaibani, S. A. Al-Mohaya, T. Sumer, E. H. Al-Sheikh, and H. Knox-Macaulay. Inherited bleeding disorders in the eastern province of saudi arabia. Acta Haematologica 1988; 79(4): 202–6.
22. Zhang L, Li H, Zhao H, Zhang X, Ji L, Yang R. Retrospective analysis of 1312 patients with hemophilia and related disorders in a single Chinese institute. Hemophilia 2003;9:696-702.
23. Manisha M, Ghosh K, Shetty S, Nair S, Khare A, Kulkarni B, et al. Spectrum of inherited bleeding disorders from Western India. Haematologia (Budap) 2002;32:39-47.
24. Sajid R, Khalid S, Mazari N, Azhar W, Khurshid M. Clinical audit of inherited bleeding disorders in a developing country. Indian J Pathol Microbiol 2010; 53:50-3.
25. Ahmad F, Kannan M, Ranjan R, Bajaj J, Choudhary P, Saxena R. Inherited platelet function disorders versus other inherited bleeding disorders: An Indian overview. Thromb Res 2008;121:835-41..



**How to cite the article:** Pallavi Mishra, Ashutosh Mishra. Study of Retrospective Analysis of Patients with Bleeding Disorders Visited in a Tertiary Care Hospital. Int J Med Res Prof. 2015, 1(2); 153-56.