

Prevalence of Hemophilia in People of Different Blood Groups in Jharkhand State: A Cross Sectional Study

Dr. Suhash Tetarway¹, **Dr. Shishir Kumar Mahto², Dr. Neha³, Dr. Sana Irfan⁴

¹. Associate Professor, Department of Physiology, Rajendra Institute of Medical Sciences, Ranchi, Jharkhand

². Tutor, Department of Physiology, Rajendra Institute of Medical Sciences, Ranchi, Jharkhand

³. Post Graduate Student, Department of Physiology, Rajendra Institute of Medical Sciences, Ranchi, Jharkhand

⁴. Post Graduate Student, Department of Physiology, Rajendra Institute of Medical Sciences, Ranchi, Jharkhand

**Corresponding author: Dr. Shishir Kumar Mahto, Tutor, Department of Physiology, Rajendra Institute of Physiology.

Abstract: Studies have shown that hemophilia, a common inherited coagulation disorder, is more common in blood group O individuals. This cross sectional study was done to evaluate the prevalence of hemophilia in people of different blood groups in Jharkhand state who are registered with the State Hemophilia Society. Main objective of this study was to evaluate the prevalence of hemophilia in patients of Jharkhand having different blood groups. Blood typing of already diagnosed cases of hemophilia A and B patients were done and prevalence of hemophilia was seen in patients of different blood groups. It was found that Prevalence of hemophilia was found to be significantly high (135 cases, n = 364) in patients with blood group O when compared with those with other blood groups and the degree of factor VIII deficiency was also high in this group of patients. The findings of this cross sectional study can be attributed to involvement of complex genetical and molecular mechanisms for which further research has to be done.

Keywords: Hemophilia, Coagulation factors, Blood groups

Date of Submission: 14-02-2020

Date of Acceptance: 29-02-2020

I. Introduction

Hemophilia is a coagulation disorder affecting approximately 1 in 1,000 men and women worldwide¹. Hemophilia A and B are single gene disorders, occurring due to a mutation in either the coagulation factor VIII or antihemophilic factor gene (hemophilia A) or the coagulation factor IX or Christmas factor gene (hemophilia B). This results into deficient synthesis of coagulation factor VIII and IX, presenting as hemorrhagic tendencies in the patients^{2, 3}. Hemophilia A is the more common variant of the disease with approximately 70 percent of case burden⁴.

Epidemiological data shows that 70 percent of hemophilia patients report positive family history and about 30 percent are sporadic cases with no report of any ethnic or geographic association⁴. Hemophilia is significantly common in India having the second highest case burden after USA⁵.

A number of studies have demonstrated a relationship between ABO blood group and hemostasis. Indeed, a higher rate of bleeding complications has been described in patients belonging to blood group O and it has also been shown that blood group O individuals are consistently overrepresented in patients with inherited bleeding disorders^{6, 7, 8}.

Furthermore, few studies have also suggested that ABO blood group is a major determinant of plasma levels of factor (FVIII) and von Willebrand factor (vWF) and blood group O individuals have significantly (approximately 25%) lower plasma levels of both these glycoproteins⁹. A clear cut correlation has been found between blood group O and levels of von Willebrand factor (vWF) and the fact has been explained with possible underlying pathophysiological mechanisms¹⁰. Recent studies have suggested that ABO blood group determinants may be important in influencing the susceptibility of plasma vWF to proteolysis by ADAMTS13 (A disintegrin and metalloproteinase with thrombospondin type 1 repeats 13) metalloproteinase¹¹. This leads to increased disintegration of vWF by proteolysis. Decreased vWF levels may in turn lead to decrease in factor VIII level as it is the specific carrier of factor VIII in plasma & protects it from proteolytic degradation, prolonging the half life of factor VIII in circulation and efficiently localizing it at the site of vascular injury¹².

So, after reviewing literature on previous studies, it is clear that there is a correlation between hemophilia and ABO blood groups and patients having blood group O are at significantly higher risk of developing hemophilia. To test these hypotheses, this study were designed to focus on to the prevalence of

hemophilia in people of different blood groups in Jharkhand state who have already reported to State Hemophilia Society of Jharkhand as no such study were conducted in the past.

Aims and Objectives

Aim of this study was to design the measures to combat the morbidity among the hemophilia patients of different blood group.

Specific objectives of the study were as follows:

1. To evaluate the prevalence of hemophilia in people of different blood groups of Jharkhand state who were registered with the State Hemophilia Society of Jharkhand.
2. Evaluation of severity and prognosis of disease in people of different blood group.

II. Materials and Methods

The present study is a cross sectional type of study done on 364 diagnosed cases of hemophilia that are registered with the State Hemophilia Society of Jharkhand. Out of them 318 patients were of hemophilia A and 46 were of hemophilia B. Proper ethical clearance was taken from the Institutional Ethics Committee (IEC), RIMS, Ranchi. Blood samples of the study subjects were obtained and their ABO and Rh typing were done. Factor assay was also done and all the data obtained were analyzed.

Observations

In this study following observations were noted:

Table: 1 (Prevalence of hemophilia in patients of different blood groups)

ABO	Rh factor	No. of Hemophilia Patients
A	Positive	92
	Negative	1
B	Positive	98
	Negative	0
AB	Positive	38
	Negative	0
O	Positive	129
	Negative	6
Total		364

In the above table, hemophilia patients have been segregated on the basis of their ABO blood groups and Rh factor. It was observed that patients with blood group O have the highest toll of hemophilia. When a comparison was done between the hemophilia patients having blood group O and that having other blood groups (A, B and AB) using Goodness of Fit test and Chi Square value was calculated, a significant difference in prevalence of hemophilia between patients with blood group O and that with other blood groups (p value < 0.00001) was observed.

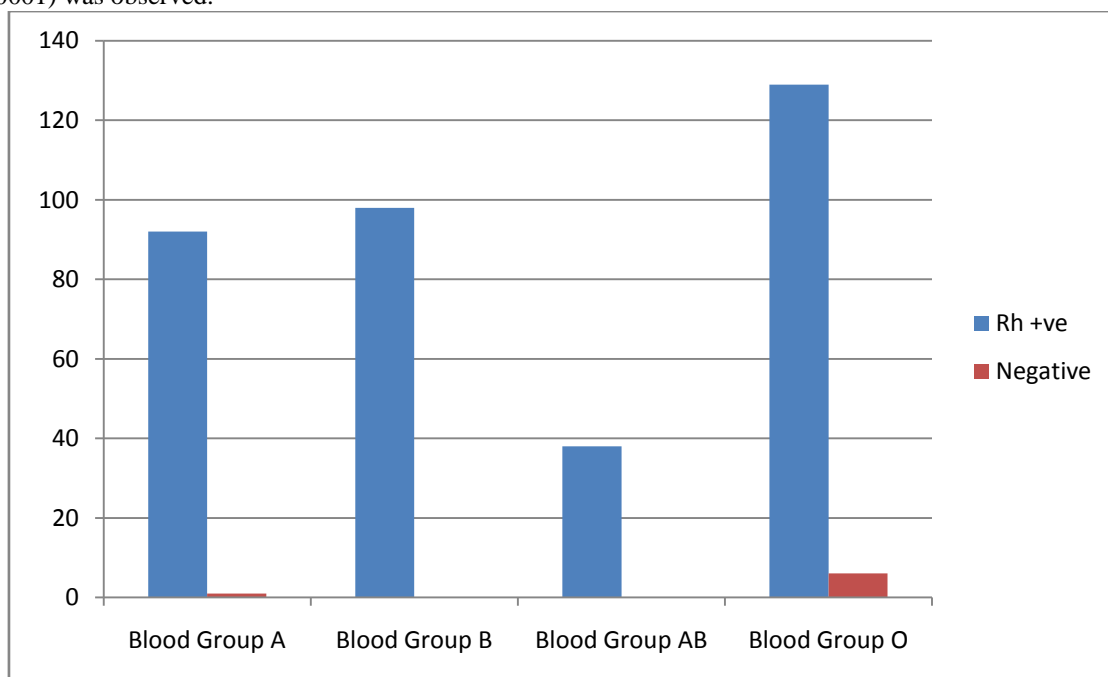
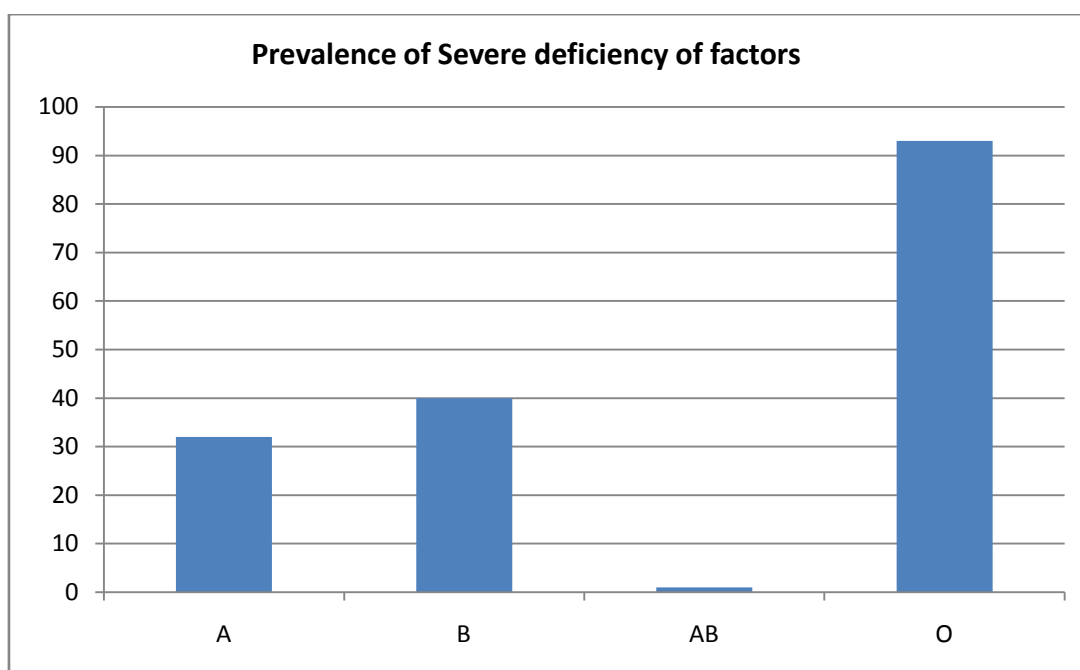


Table: 2 (Degree of factor deficiency in hemophilia patients with different blood groups)¹³

Degree of deficiency of factors	Blood Group				Total
	A	B	AB	O	
Mild (Factor level 5 – 40% of normal)	38	32	31	22	123
Moderate (Factor level 1 – 5% of normal)	23	26	6	20	75
Severe (Factor level < 1% of normal)	32	40	1	93	161

In this table, hemophilia patients with different blood groups were classified on the basis of their degree of coagulation factor deficiency. It was observed that severity of factor deficiency is more in blood group O and it is less in blood group AB.



III. Conclusion

In our study it was concluded that hemophilia was significantly higher in blood group O and the degree of factor deficiency was severe (i.e. less than 1% of normal) in these patients. These findings clearly indicate that blood group O somehow causes decrease in plasma levels of coagulation factors VIII and IX leading to development of hemophilia. Some complex genetic and molecular mechanisms may be involved in causation of these events which are needed to be studied further

Severity of coagulation factor deficiency is more in blood group O whereas it is less in blood group AB. This fact needs further evaluation in order to understand the pathogenesis and underlying genetic and molecular bases or variation in development of this crippling disorder.

IV. Discussion

The findings of this cross sectional study suggest that blood group O hemophilic individuals should be paid special attention in terms of their treatment and care. As the degree of factor deficiency is significantly higher in blood group O individuals, they are always prone to succumb into acute bleeding tendencies. They must be educated and counseled regarding these facts strategies should also be made to combat any emergency situation, like adequate arrangements of factors in hemophilia treatment centres and maintenance of adequate stock of group O blood and other blood products in blood banks so that their morbidity and mortality can be reduced. Also, prior to any surgery to this category of hemophilic patients, special attention must be paid towards their plasma levels of factors and inhibitors.

Furthermore, the genetic and molecular bases of the fact of increased susceptibility of blood group O individuals to hemophilia must be studied and this will open up doors of new research arena. The research must also be done to study the probability of development of inhibitors in this group of hemophilic patients.

References:

- [1]. Skinner MW. WFH: closing the global gap - achieving optimal care. *Haemophilia*. 2012; 18 (Suppl 4):1–12. [PubMed]
- [2]. Kemball-Cook G, Gomez K. Molecular basis of hemophilia A. In: Lee C, Berntorp E, Hoots K, editors. *Textbook of hemophilia*. 2nd ed. West Sussex: Wiley-Blackwell; 2010. pp. 24–32.
- [3]. Gomez K. Hemophilia B-molecular basis. In: Lee C, Berntorp E, Hoots K, editors. *Textbook of hemophilia*. 2nd ed. West Sussex: Wiley-Blackwell; 2010. pp. 88–93.
- [4]. Haldane JB. The rate of spontaneous mutation of a human gene. *J Genet*. 1935; 31:317–26. [PubMed]
- [5]. World Federation of Hemophilia (WFH) Canada: WFH; 2013. [Accessed on May 9, 2013]. Report on the Annual Global Survey 2011. Available from: <http://www1.wfh.org/publications/files/pdf-1488.pdf>
- [6]. Gill JC, Endres-Brooks J, Bauer PJ, Marks WJ, Jr, Montgomery RR. The effect of ABO blood group on the diagnosis of von Willebrand disease. *Blood*. 1987; 69:1691–1695. [PubMed]
- [7]. Horwich L, Evans D, McConnell R, Donohoe W. ABO blood groups in gastric bleeding. *Gut*. 1966; 7:680–685. doi: 10.1136/gut.7.6.680. [PMC free article] [PubMed] [CrossRef]
- [8]. Evans D, Horwich L, McConnell R, Bullen M. Influence of the ABO blood groups and secretor status on bleeding and on perforation of duodenal ulcer. *Gut*. 1968; 9:319–322. doi: 10.1136/gut.9.3.319. [PMC free article] [PubMed] [CrossRef]
- [9]. O'Donnell J, Laffan MA. The relationship between ABO histo-blood group, factor VIII and von Willebrand factor. *Transfus Med*. 2001 Aug; 11(4):343-51.
- [10]. Massimo Franchini, Franco Capra, Giovanni Targher, Martina Montagnana, and Giuseppe Lippi. Relationship between ABO blood group and von Willebrand factor levels: from biology to clinical implications. *Thromb J*. 2007; 5: 14. Published online 2007 Sep 25. doi: 10.1186/1477-9560-5-14 PMID: 17894864
- [11]. Jenkins PV, O'Donnell JS. ABO blood group determines plasma von Willebrand factor levels: a biologic function after all? *Transfusion*. 2006;46: 1836–1844. doi: 10.1111/j.1537-2995.2006.00975.x. [PubMed] [CrossRef]
- [12]. Vlot AJ, Koppelman SJ, Bouma BN, Sixma JJ. Factor VIII and von Willebrand Factor. *Thromb Haemost*. 1998; 79:46–465. [PubMed]
- [13]. <https://emedicine.medscape.com/article/779322-overview>

Dr. Shishir Kumar Mahto, et al. "Prevalence of Hemophilia in People of Different Blood Groups in Jharkhand State: A Cross Sectional Study." *IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)*, 19(2), 2020, pp. 33-36.