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Clinical, Demographic, And Haematological Profile of Haemophilia Patients at Sir Sunder Lal Hospital Varanasi With CoE Haemoglobinopathies And Day Care Centre

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Abstract

Introduction: Hemophilia is a recessive coagulation and bleeding disorder that is X-linked and inheritable. The Factor VIII (F8) and Factor IX (F9) genes are involved in hemophilia A and B, two hereditary diseases. The process of hemostasis is hampered by spontaneous or post-traumatic bleeding, which also increases the risk of hemophilia. Aim: The purpose of our study is to evaluate the hematological, clinical, and demographic traits of individuals with hemophilia A and B.

Results: A total of 646 hemophilia patients were examined during the study period. the majority of hemophilia A patient 544 (84.21%) and hemophilia B 102 (16.01%). Based on severity, severe 342 (52.94%), Moderate 247 (38.24%), and Mild (8.82%) had been diagnosed Most common age group was <1 -58 years. The bulk of the study population had a mean age of (Mean ± SD=18.45±13.00) and a median of 17. Maximum number of patients at the ages of 21 and 30 years (21.05%) and a median of 25. The most common age of onset is <1 year. On the other hand, 61.3% of patients had hemophilia when they received their initial diagnosis. The median number of bouts of

spontaneous bleeding across all patients was 17 and there was no discernible difference between those with moderate and severe symptoms of bleeding. Hemarthrosis infection (target joint) was most prevalent (79.10%), then prolonged bleeding after cut/ trauma (62.38%). Hemarthrosis knee joint was the joint that was infected (target joint) the most common (79.10%), followed by joint bleeding (46.75%) Severe disease affected the knee joint more frequently than moderate disease.

Conclusion: The present research revealed differences in the incidence and nature of bleeding on its own in hemophilia patients. Gum bleeding and post-traumatic bleeding in a child who is otherwise healthy should alert the doctor to consider hemophilia.

Keywords: Hemophilia A, Hemophilia B Factor VIII, Factor IX

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I. Background

Hemophilia is an X-linked recessive congenital bleeding disorder. Haemophilia A (Classical) and B (Christmas sickness) are brought on caused, respectively, by a lack of coagulation factors VIII (FVIII) or IX (FIX). Haemophilia A and B account for 80% and 20% of all instances of hemophilia, respectively [1]. Most inherited clotting problems are represented by it. Hemophilia A and B affect roughly one in every 5,000 and one in every 30,000 male newborns, respectively **[2].** However, over 39.78% of cases are caused by de novo mutations and have no family history, and 60.31% of cases present positive family history **[3**]. 1,125,000 people worldwide suffer from hemophilia **[4]**, 80% of them are in underdeveloped nations **[3].** India donates a substantial percentage of the patient burden. According to the "World Hemophilia Organization," there are 1675 registered patients. Registry for Histoplasma Haemophilia affects 10 per million people in this country **[5],** while the true prevalence may be significantly higher due to survey method limitations that prevent all patients from being assessed and monitored consistently. Based on the remaining endogenous FVIII/FIX concentrations, hemophilia patients are grouped. Patients with hemophilia are categorized on the levels of endogenous FVIII/FIX that are still present. The severity of hemophilia in participants with factor levels of greater than 1, greater than 1 to 5, and greater than 5 IU/dL was assessed. Hemophilia patients make up about half of the population disease. But even in cases of severe sickness, individuals are seen with uneven bleeding episodes. **[6]**. Hemophilia's categorization provides information on the

likelihood and frequency of recurrent bleeding. Hemophiliacs who have severe spontaneous bleeding may do so after minor to moderate traumas, while those who have a moderate form may experience mild to moderate spontaneous bleeding. Mild hemophilia patients can go years before being diagnosed, and they only experience bleeding after severe wounds or major surgery **[7].**

Hemarthrosis is the most common complication and the tell-tale sign of a severe case of hemophilia [8–9]. Either naturally or following trauma, it happens. More than 45% of hemophilic children in Bangladesh were observed to have joint complaints, particularly in the knees. The most common type is joint arthropathy **[1].** However, a majority of ankle joints were involved, according to several European research **[10, 11].** Muscular bleeding is regarded as one of the main causes of impairment in this condition and It affects 10–25% of severe hemophilia hemorrhagic episodes **[7]**. Muscle bleeding affects about three-quarters of people with severe hemophilia time. A potentially fatal hemorrhage called an iliopsoas hematoma has been shown to potentially compress the femoral nerve. An additional factor in hemophilia-related morbidity and mortality CNS bleeding exists **[8].** Hemophilia occasionally results in intestinal hemorrhage. It could happen haphazardly or due to typical gastrointestinal bleeding causes. Another bothersome symptom is hematuria ureteral blockage or hydronephrosis in some people may be complicated by hemophilia **[7].**

Investigating In some cases, the incidence and locations of bleeding in Hemophilics could aid in the early diagnosis and prognosis of hemophilia. that have not yet received a diagnosis or suspicion. By extension, it enables timely safeguards. Consequently, the goal of the current investigation was to elucidate the demographics and clinical manifestations of patients who have mild, moderate, or severe hemophilia, closely monitoring the sites and patterns of bleeding.

II. Introduction:

The most frequent inherited conditions are factors VIII (F8) and IX (F9) deficiency. There are several bleeding illnesses, and patients might appear with signs of bleeding in any area of the hospital. Therefore, details concerning Prevalence, clinical characteristics, and research are crucial for Hemophilia treatment and diagnosis. The diagnosis is carried out in coagulation laboratories, which involves specialist testing because there are few laboratories. These specialist examinations can be performed department of pathology. The goal of this study was to fill this informational gap by calculating the proportion of individuals with hemophilia who are referred to hemophilia day care units and examining the clinic-pathological characteristics of these sufferers at this tertiary care facility.[1]

The clinicopathological feature of hemophilia patients is hemorrhage in muscles, joints, and soft tissues. A hemophilia patient's factor levels 1% and 5% (0.01-0.05 IU/ML), less than 1% (0.01 IU/ML), or higher, result in severe, moderate, or mild hemophilia and above 5% to 40% (>0.05-0.40 IU/ML) (normal values = 50–150%)[2]. Patients with severe, moderate, and Mild hemophilia bleeding occur spontaneously or with minor cuts and trauma, Generally Moderate and Mild bleeding after major surgery and significant trauma. In underdeveloped countries like India, Limited treatment options are available for those with hemophilia, frequent joint bleeds cause significant disability, and joint impairment morbidity rises significantly with age. [3]additionally, The probability of infections arises when blood and blood products are often used instead of protein concentrate because they are less expensive to transfer during transfusions.

Deficiency in factors VIII and IX (A and B bleeding disorders, respectively).

Defects in Von Willebrand's factor and factor B) are the two most common coagulation factors. Additional coagulation factors that are hereditarily deficient include Significantly less frequent.[3] In local or ethnic populations Recessively inherited in areas where consanguineous couplings are widespread More people experience bleeding issues. Haemophilia A is present in one out of every 10,000 male babies, and one in every 30,000 male births has hemophilia B. Haemophilia is prevalent, and Depending on the reporting country, A ranges from 5.4 to 14.5 incidences per 100 thousand males.[4,5]

III. Objective:

To evaluate the prevalence, clinical, demographic, and hematological characteristics of hereditary coagulation diseases, specifically hemophilia A and B.

IV. Material & Methods:

The Sir Sunder Lal Hospital Institute of Medical Science BHU conducted this in vestigation.

From March 2021 to August 2024 following an evaluation of the clinical profile, age of the first hemorrhage age of the initial diagnosis, family history, age of the first joint bleed, and bleeding history from the previous year, the type of treatment utilized, the frequency of bleeding in the most affected joint, and the type of treatment. Way of ancestry, a history of previous treatment, and blood thorough physical examination were done in addition to the transfusion. These tests included thermostatic Prothrombin time (PT), General Blood Pressure (GBP), Complete Blood Count (CBC), Activated Partial Thromboplastin Time (APTT), and an Investigation to Address the Deficiency of a Particular Factor Regarding the Presence of Al(OH)3 Adsorbed Plasma, Regular Aged Serum, and a Specific Factor Assay test within 4 hours of the

blood sample, all tests were run. However after one or two days, factor VIII assays were conducted since the plasma was stored at a temperature between 20 and 30 °C. Factor VIII is measured using the Bethesda assay. A source is combined with test plasma. Assays for factors VIII and IX were performed on all new cases (if not already completed). In earlier circumstances, factor level was only confirmed when it was done within 24 hours of getting blood or the factor VIII/IX method used wasn't a one-stage assay. Semi-automated clot analysis was used for the "one-stage assay" analyzer. The basis for this is an assessment of dilutions' capacity to reverse the plasma's activated partial thromboplastin time, which is wholly useless in F VIII, but also has every other component needed for clotting. Factor values of less than 0.1 to 0.05 IU/mL (1 to 5%), less than 0.01 IU/mL (<1%), and Significant, moderate, severe, and mild hemophilia are defined as >0.05-0.40 IU/mL (>5-<40%) respectively. Additionally, all cases underwent testing for HIV, hepatitis B, and hepatitis C. One patient is diagnosed as HIV positive.

Statistical Analysis:

Using qualitative variable descriptive analysis, percentages, and frequency. The mean value of the statistical average was calculated, and the standard deviation was used to measure of dispersion.

V. Results:

This study was conducted at CoE Haemoglobinopathies and Day Care Centre at a tertiary care hospital Sir Sunder Lal Hospital IMS BHU. There were 646 patients studied in all during the duration of the study we were conducting. Out of which 544 (84.21%) patients were of hemophilia A and 102 (15.81%) patients of hemophilia B **[Table no. 1] [Figure No.1A].**

Patient distribution based on hemophilia type		
Types of hemophilia	Number of patients	Percentage (%)
Hemophilia A	544	84.21
Hemophilia B	102	15.81
Total	646	100%

Table no.1 Patient distribution based on hemophilia type

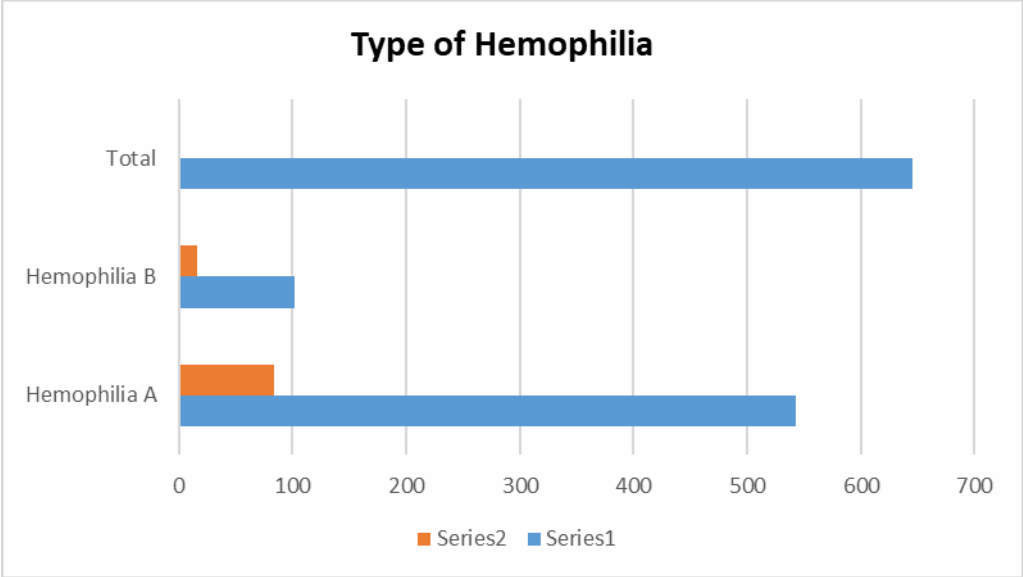

Figure No.1. A. Shows types of hemophilia

The overall age distribution of hemophilia patients' mean age and standard deviation is (Mean±SD 18.45±13.00). The majority of cases (21.05%) in the study group were in the 21–30year age range (Mean±SD24.65±2.85), followed by 17.33% (Mean ±SD 18.04 ±1.30) in the 16–20year range, 6-10year age range and 11-15year age range is 15.59% (mean ± SD 8.05±1.45) and (Mean±SD13.02±1.41), 14.7.% > 30-year age range (Mean ± SD 42.26±10.57), and the study group's average age was 18.45 13.00 years. Five patients (0.77%) had the commencement of their symptoms before one month, however, none were identified during that time. Similarly, although the majority (92%) of children exhibited symptoms by the time they turned 5 years old, only 76% had received a diagnosis **[Table No.2].**

	Age distribution of patient			
Serial Number	**Year group of age**	**Male**	**Female**	**Percentage(%)**
1	<1	5	-	0.77%
2	1-5	90	-	13.93%
3	6-10	103	-	15.95%
4	11-15	104	--	16.10%
5	16-20	112	-	17.33%
6	21-30	136	1	21.05%
7	>31	95	-	14.70%
Age(Mean±SD)	18.45±13.00			
Total		646		100%

Table. no. 2. Shows the age Distribution of patients

Family history was advantageous in 341 of 544 hemophilia A patients (63%) and 48 in 102 hemophilia B (47%). Only one female is found positive family history [**Table no.3.A**]. **[Figure No.1.B.]**. 103 out of 544 hemophilia A and 9 in 102 found consanguinity. History of plasma transfusions occurs in 401 out of 544 (74%) in hemophilia A and 54 in 102 (53%) in hemophilia B. One patient of hemophilia A found HIV positive and the rest of them gave negative hepatitis B, Hepatitis C, and HIV tests.

Patients' Distribution with a Positive Family Background (n=645)				
Family History	Hemophilia A (n= 544)	Hemophilia B (n= 102)	Total	Percentage (%)
Present	341	48	389	60.31%
Absent	203	54	257	39.78%
Total	544	102	646	100%

Table no. 3. A. Shows Patients' Distribution with a Positive Family Background

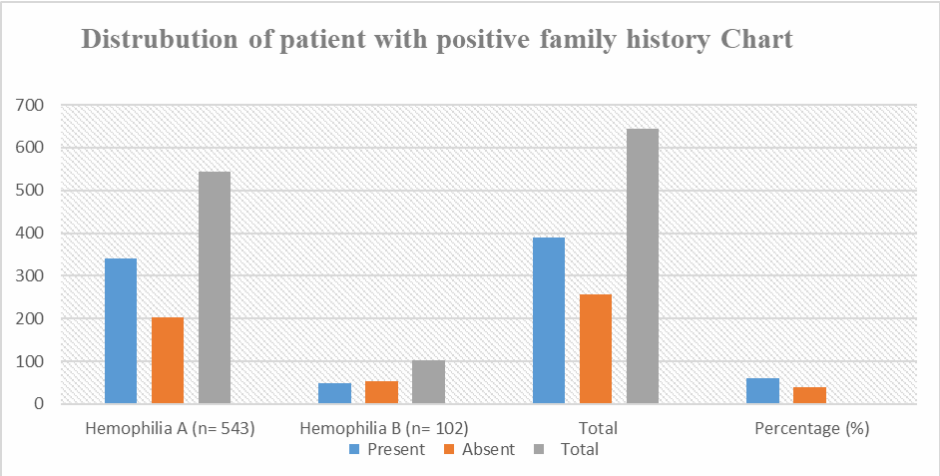

Figure No. 1.B. Shows Distribution of patients with positive family history

Based on severity hemophilia A and B are categorized as mild (>5-<40% or >0.05-0.40 IU/mL), moderate (1-5% or of <0.1 to 0.05 IU/mL), and severe (<1% or < 0.01 IU/mL), (normal factor level 50-150%). Clinical manifestations of patients according to the severity in hemophilia 342 (52.94%) severe, moderate 247 (38.24%), and mild 57 (8.82%) shown in **[Table no.3. B.] [Figure no.1. C.].** All cases of hemophilia A and B had prolonged bleeding following trauma, according to the patient's clinical signs and symptoms. **[Table No. S1].** Frequency and

Distribution of Haemarthrosis in Hemophilia A and Hemophilia B **[Table No. S2].**

Clinical Manifestation of Patient According to severity (n= 646)		
Severity of Hemophilia	Number of patients	Percentage (%)
Severe Hemophilia	342	52.94%
Moderate Hemophilia	247	38.24%
Mild Hemophilia	57	8.82%
	646	100.00%

Table No. 3.B. Clinical Manifestation of Patient According to Severity

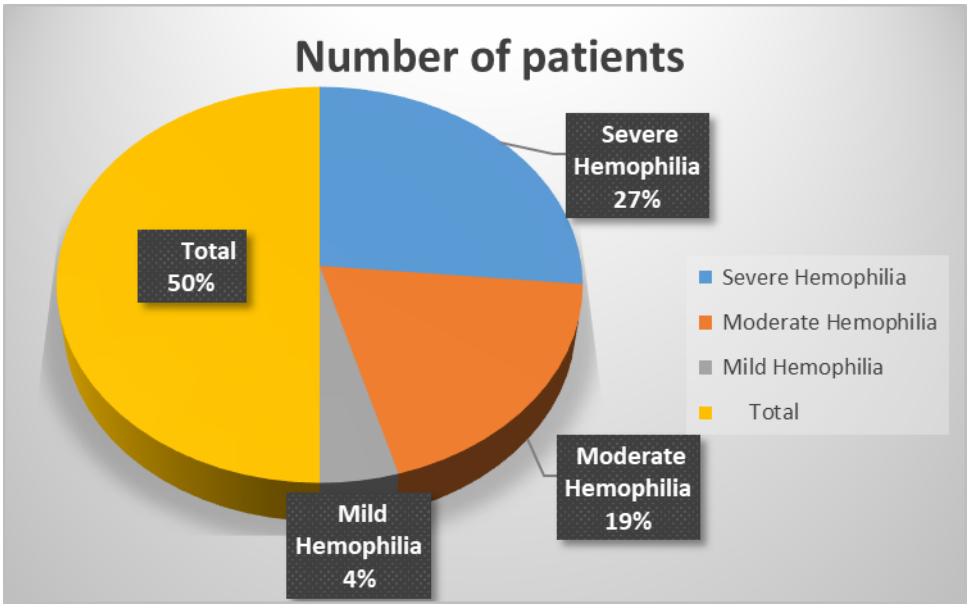

Figure No. 1. C. Shows the number of hemophilia patient

	Laboratory Presentation in Haemophilia Patients					
	Haemophilia A and Hemophilia B					
	severe	moderate	Mild	severe	Moderate	Mild
PT control (mean)	13.1-13.5 (13.3)s	13-13.5 (13.3s)	13-13.5 (13.3)s	13.1-13.5 (13.3) s	13.0-13.4 (13.3) s	13.0-13.6(13.3)s
PT test (mean)	12.7-16.5 (13.9) s	12.9-16 (13.9) s	13-15.5 (13.5) s	13-17 (13.7) s	12.4-13.7 (13) s	13.5-13.6 s
APTT control (mean)	28-30 (29)s	28-30 (29)s	28-30 (29)s	28-30s (29)s	28-30 (29)s	28-30 (29)s
APTT test	67.9-122	47.1-105	42-63.1(52)	69-136 (107)s	62- 64 (63)s	44-57 (50.5)s

(mean)	(90)s	(68)s	s			
FVIII	0.2-.99 (0.6) s	1-5 (2.3)s	5.5-17 (16)s	Normal	Normal	Normal
FVIII control	86-132 (109)	85- 132 (108.5)	85-121(103)s	Normal	Normal	Normal
FXI	Normal	Normal	Normal	0.2-0.97 (0.6)	1.3-3.0 (2.2)	7-15.9 (11.5)
FIX control	Normal	Normal	Normal	90-132 (111)	85-121 (103)	109-115(107)
Platelets	**Adequate**	**Adequate**	**Adequate**	**Adequate**	**Adequate**	**Adequate**

Table No. 3. C. Shows the Laboratory Presentation in Haemophilia Patients

The majority of cases in both hemophilia A and B. APTT increased in all patients usually 67 seconds. According to [**Table No. 3. C.],** every type of bleeding was present in every patient. The most often targeted joint was knee hemarthrosis (79.10%), followed by prolonged bleeding on cut and trauma (62.38%). 55.57% of patients had gum bleeding, 47.83% had Echymosis /bruise, joint bleeds 46.75%, hematemesis 33.44% stool bleeds 33.28%, and hematoma 44.74%. However, there was not a significant distinction in any of the presenting symptoms between moderate and severe hemophilia.

Overall, 65.9% of patients displayed muscle hematoma ultrasonographic characteristics, the entire abdomen's USG revealed retroperitoneal hemorrhage in 27.9% of patients., and 93.0% revealed alterations of the infected joint on X-ray. Table No. 5 and Fig. 3 display the Participants with hemophilia who participated in the study's position and the incidence of unexpected bleeding about severity. For severe, moderate, and complete hemophiliac individuals, respectively, the median number of spontaneous bleeding episodes during the past two years was 40, 25, and 19, with a range of 0 to 97 occurrences overall. The knee joint was the joint that was infected the most (79.10%), subsequently followed by the ankle and elbow joints (46.75%). Knee joints had the largest median number of incidents of joint bleeding (14, range: 1-40). Elbow joints were next, with a median of 10 occurrences per episode and a range of 1 to 32. Ankle, elbow, and knee bleeding were more frequent in severe hemophiliacs, and Moderate hemophiliacs were more likely to experience shoulder joint hemorrhage. In contrast, the difference was not statistically significant.

The thigh muscle was the most variable concerning range, frequency, and number of episodes. the most often affected muscle group among mild hemophiliacs compared to severe hemophiliacs. The difference, once more, was not statistically significant. Both categories of patients had gum bleeding, with

bleeding episodes being more frequent in the severe group. 33.44% of patients had hematemesis. Both the moderate and severe categories showed a similar distribution. Both patient groups experienced almost identical amounts of genitourinary hemorrhage.

There was no sign of cerebral bleeding or retinal bleeding. Paradoxically, only the frequency of bruise and ecchymosis episodes significantly differed between moderate and severe hemophilia patients.

VI. Discussion:

In the current investigation, 646 patients with atypical bleeding symptoms were examined. Out of which 544 (84.21%) individuals had hemophilia A, and 102 (15.81%) had hemophilia B. (Table no. 1). Hemophilia A and B Coagulation Bleeding disorder. More individuals have hemophilia A than hemophilia B (Christmas disease). The condition could manifest at any age between 2.5 months and 54 years. Out of these, 16.40% (106/646) of the patients are children (0–18 years), and 83.59% (540/646) of the patients are adults (19–58 years). A maximum number of patients 32.50%% (210/646) were between the ages of 21 and 30; the next group was composed of those between the ages of 16 and 20. 31.89% (206/646) >1 five years. The lowest possible number of patients is a part of the 1–5year age category, with 1.39% (9/646) > 30 years (16.72%)> Less than a year (6.34%) > 6 to 10 years. The patients' ages ranged from 11.25 to +8.84 years, with a 10-year median and a 15-year mean. varied from 2 to 47 years, with the average being 32.2 years median age at manifestation, according to research by Gupta and colleagues. The age range at manifestation, according to Sajid et al.(7), was 3 to 57 years of age, with 17 as the median age. The majority of patients have a mild diagnostic of an adult group deficit. 15.8 years old as the mean was observed by Munira et al.9 There were only male patients in the present study. except one patient is female is diagnosed with hemophilia A.

In the current investigation, there were only male patients. Given that X-linked hereditary coagulated bleeding disorders comprise patients suffering hemophilia A and B are more likely to have it while females serve as carriers. In the current investigation, persistent bleeding after a cut was the most frequent presenting clinical characteristic (79.10%)., > haemarthrosis (41.30%), >gum bleeding (55.57%), >Ecchymosis/bruise (47.83%), >joint bleeds (46.75%), >Haematoma (44.74%), >hematemesis (33.44%), >Hematuria (31.73%), >Petechiae (22.45%), >skin bleeds (16.40%), > muscle bleeds (15.33%), > posttraumatic bleeds (15.17%), > Epistaxis (10.68%), and tooth extraction (3.25%), bleeding following a tonsillectomy (3.26%), bleeding following a circumcision (2.32%), and bleeding after Cephalohematoma **(Table No. S1).**

Hemarthrosis (82%) was the most prevalent presenting characteristic in hemophilia, according to Ahmed et al. [10] According to Munira Borhany et al.,

haemarthrosis occurred in 72.8 percent of cases, hematoma in 51.4%, post-circumcision bleeding in 37.14 percent, and bleeding after trauma in 28.51 percent, with haematuria, bruises, and gum bleeding following. [9] In the current study, 40.50 percent of participants reported having at least one instance of joint swelling involving one, two, or more knee joints being the most prevalent. The most common joint involvement, according to Sajid et al., is the knee joint (48%), while in 36% of cases, more than one joint is involved. is implicated. [8] 1% of factor VIIIc categorized as severe hemophilia A was present in 52.94% (342/646) patients, while 15% of factor VIIIc was present in 38.24% (247/646) instances moderate hemophilia A, while 8.82% (57/646) had > 5% factor VIIIc classified as mild hemophilia A.

77.8% of severe hemophilia A patients were found in research by Ahmed et al. 7.75% of instances of mild hemophilia A and 14.4% of cases of moderate hemophilia A were recorded. [10] Out of 212 Haemophilia A individuals37.2% (79) moderate, 41% (87) intermediate, and 21.6% (46) severe cases of hemophilia were described by Sajid et al. **[8].** Positive findings were obtained in research by Saurabh Mishra 13 et al. (64.96%), Raina 14 et al. (75%), R.K. Nigam 12 et al. (76.7%), Karim et al. 11 (82%), and Shamoon 15 et al. (68.57%). according to current studies, knee Based on research conducted by MA The knee joint was the subject of studies by Karim 17 et al., RK Nigam 14 et al. (knee joint - 64.96%), Saurabh Mishra 18 et al. (knee joint - 57%), and Payal 14 et al. (knee joint - 58%). Knee joint: 61.43% according to Raina23 et al, Shamoon et al[24], and 67.8%). (60.1% of knee joints.) 52% of participants in the current study have moderate to severe hemophilia affecting 22% of people. moderate hemophilia, which affects 26% of people percentage of mild, moderate, and severe patients. The study's description of hemophilia corresponds to one by Sadaria[24] et al.

The clinical presentation of the patients and these statistics are comparable. Patients with very low concentrations of factor VIIIc visited clinics because their symptoms were so severe, whereas he symptoms of patients with mild to severe factor VIIIc insufficiency could be managed locally, they did not visit clinics. Hemophilia is a condition that affects the joints, making it difficult to carry out daily tasks and changing how one functions in social situations. Due to the availability of focuses on safety factors and thoroughly multidisciplinary comprehensive care approaches, hemophiliacs in developed nations enjoy a degree of quality of life that is quite similar to what is experienced by people of all ages. However, in developing nations like ours, due to cost-related concerns, because safe products and acceptable product attributes are not used to treat hemophiliacs, this might result in pain, arthropathy, and impairment.

Even though the present situation, hemarthrosis was the most prevalent hemophilia-presenting symptom in children, and bruising and hematomas,

either traumatizing or spontaneous, were the primary symptoms at the time of the demonstration made for these children. Therefore, a youngster who is generally healthy should be evaluated for hemophilia if they exhibit these characteristics. Hemophilia patients may experience a variety of clinical symptoms and require continuing care. The initial for the time being, Primary care physicians continue to be the primary care providers.
Moderate to mild symptoms that can be treated at home or in a hospital. The Center is run by a primary care doctor. This investigation will support the frequency, clinical characteristics, and laboratory data diagnostic of hemophilia so that medical professionals can make a diagnosis. They specifically take care of patients with hemophilia and hereditary bleeding disorders. A diagnosis is necessary before beginning the management of the sufferers. Patients with hemophilia may experience a variety of clinical symptoms and require continuing treatment. the original Primary care doctors continue to be the point of contact and care. Along with the identification and management of mild to moderate Ina daycare or at home, symptoms can be managed in a primary care physician-run facility. This research will assist the frequency, clinical features, and lab data for the purpose of allowing medical experts to diagnose hemophilia Patients with inherited bleeding disorders are treated particularly by them hemophilia. Before starting management, a diagnosis is required of the patients.

VII. Conclusion:

Depending on the severity of the disease, hemophiliacs can appear in a variety of ways. Blood loss is frequently and persistently related even with minor trauma, particularly when there is bleeding, bruises, and hemorrhages that are either spontaneous or trauma in an otherwise healthy youngster should raise concerns. even in the absence of hemophilia, a doctor should check the patient of kinship history to understand the range of presenting styles a population's awareness of hemophilia aids in early diagnosis and management strategy. Primary care physicians play a crucial role in delivering routine care, treating bleeding emergencies, and monitoring patients for anemia caused by hemorrhage, transfusion-transmitted infections, and joint deformities. Additionally, doctors can lower morbidity and mortality associated with severe hemophilia by administering clotting factors as a preventative measure. Even though gene therapy and other recent advancements in therapy offer hope for the future, this region of India still has a great need for diagnostic and treatment facilities.

Ethics approval and consent to participate- The Institute of Medical Science, Banaras Hindu University, obtained institutional ethical permission (No: Dean/2022/EC/3611) from the ethical committee.

Consent for publication- The patient or participant provided written informed consent for publication.

Availability of data and material- The corresponding author can provide the datasets used and/or analyzed in the current work upon reasonable request.

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