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TO STUDY PATTERN OF JOINT INVOLVEMENT IN PERSONS WITH HEMOPHILIA AT TERTIARY CARE HAMIDIA HOSPITAL, GANDHI MEDICAL COLLEGE, BHOPAL.

Pathology

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ABSTRACT

Introduction: The X-linked inherited coagulation disorders, hemophilia-A (Factor VIII deficiency) and hemophilia-B(factor IX deficiency), together with Von Willebrand disease comprise 95 to 97 percent of all the Inherited Deficiencies of coagulation factors.[1,2] **Aim:** The present study was conducted with the aim to study pattern of joint involvement in Persons with Hemophilia (PWH), at Hemophilia Day Care Center G.M.C. Bhopal. **Settings And Designs:** The retrospective hospital-based study **Materials And Methods:** All the patients diagnosed with Hemophilia-A, Hemophilia-B & Von willebrand disease, on the basis of history, physical examinations & laboratory investigations at Hemophilia Day Care Centre & Department of Pathology G.M.C.Bhopal. **Results:** A record of total of 579 PWH were collected, out of which 511 (88.2%)cases were hemophilia-A where most common presenting complaint was joint bleeding 404 (79%) cases, out of which bilateral knee joint 38(9.4%) cases were found to be predominantly involved. 57(9.8%) cases were diagnosed, Hemophilia-B with the presenting complaint of joint bleeding 41(71.9%)cases out of which bilateral knee 09 (21.1%)cases involved was most commonly. Total 11 (1.8%) cases of Von Willebrand disease with none having joint involvement and most common presenting complaint was abdominal bleeding 08(72%)cases, in the all three disorders other symptoms were petechiae, epistaxis, bleeding gum and muscle hematoma.**Conclusion:** Bleeding after injury is obvious in healthy people but in severe Hemophilic bleeding can occur even without trauma. Hemarthrosis was the most prevalent consequence & knee joint was most common affected joint in PWH. In a healthy child, the presence of bruises, hematomas & hemarthrosis should prompt further examinations into disorder particularly hemophilia.

KEYWORDS

Hemarthrosis, bruises, hematoma

Introduction

Patients with hemophilia frequently experience joint involvement, resulting in significant morbidity from arthropathy. Hemophilia-A (Factor VIII deficiency), hemophilia-B (factor IX deficiency) & Von-Willebrand disease comprise 95% to 97% of all the Inherited deficiencies of coagulation factors.[1,2] Inadequate FVIII levels result in the insufficient formation of the intrinsic tenase complex (FIXa, FVIIIa, Ca2+ and Phospholipids) which subsequently leads to reduced generation of thrombin with impaired fibrin deposition and clot formation.[3] Haemophilia usually occurs in males, whereas females are generally carriers.[4] The prevalence of Hemophilia-A is 1 in 10,000 and that of Hemophilia-B is 1 in 30,000-50,000.[5,6]Male live births and Von- Willebrand disease affecting nearly 1% of population.[6] Patients with severe haemophilia frequently bleed into their muscles or joint.

Hemarthrosis is the most common, the most painful and the most physically, economically and psychologically debilitating manifestation of the hemophilia A.[7,8] Spontaneous bleeding is uncommon in people with moderate haemophilia. Mild haemophilia patients typically bleed as a result of major surgery or injury. They do not bleed often, and some may never have a bleeding problem.[9] Replacement of the deficient factor is the mainstay of treatment however, if factor concentrates are either not available or not affordable, transfusion options including whole blood, FFP (fresh frozen plasma) & Cryoprecipitate are still being used. Minor bleeds have also been controlled with antifibrinolytics agents.[10]

MATERIALS AND METHODS-

This retrospective study was conducted in the Department of Pathology in collaboration with Hemophilia Day Care Centre at Gandhi Medical College, Bhopal during the period of January 2021 to December 2021.

A total 579 persons with coagulation disorder visiting for various laboratory tests with different medical and surgical complaints finally diagnosed as hemophilia were included in this study. A thorough history including family history, findings of physical examination & laboratory tests were taken from medical records. Diagnosis was made on the basis of history, physical examination and laboratory investigations such as bleeding time (BT), Prothrombin time (PT), Activated Partial Thromboplastin Time (APTT), Mixing & correction studies (factor VIII and factor IX estimation). Other investigations like complete blood counts including peripheral blood smear were also made to see blood cell morphology and platelet count & blood grouping. Results of the investigations were recorded and analyzed using MS excel.

Exclusion Criterion:

Patients with platelet disorders were excluded.

Ethical Approved:

Approved by Hemophilia Society in G.M.C. Bhopal.

Funding: No funding sources.

RESULTS-

In the over study 579 cases attending with history of prolonged bleeding along with various medical and surgical complaints, finally diagnosed as Hemophilia A& B and Von Willebrand diseases in the Department of Pathology GMC & Hamidia Hospital Bhopal were included. A total of 579 cases of hemophilia were found, out of which 511(88.2%), 57(9.8%) & 11(1.8) Cases were Hemophilia-A, Hemophilia-B & von willebrand disease respectively.. Further analysis of 579 hemophilia patients was done.

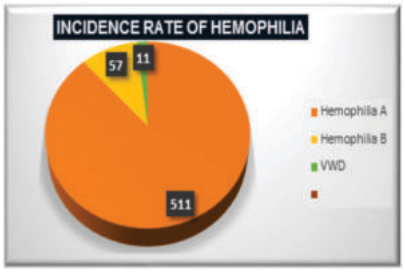
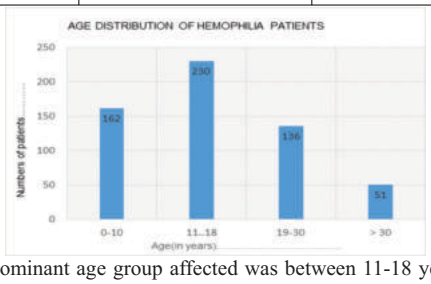


Table 1: Age Distribution Of The Hemophilia Patients

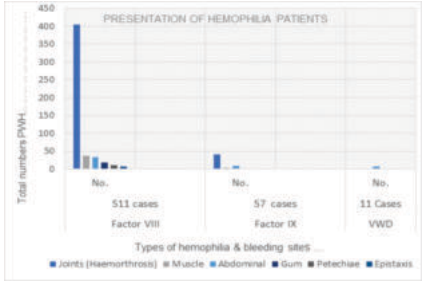
Age (in years)	No. of Patients	Percentage
0-10	162	27.97%
11-18	230	39.72%
19-30	136	23.48%
> 30	51	8.80%
Total no.	579	



The predominant age group affected was between 11-18 years- 230 (39.72%) (Table 1) Although they were ranging from 8months to 42 years.

Table 2. 1st Presentation Of Hemophilia Patients

	Factor VIII		Factor IX		VWD		Total No.
	No.	%	No.	%	No	%	
Joints	404	69.77	41	7.08	00	00	445
muscles	37	6.39	03	0.51	01	0.17	41
Abdominal	34	5.87	09	1.55	08	1.38	51
Gum	18	3.10	01	0.17	01	0.17	20
Petechiae	10	1.7	02	0.34	01	0.17	13
Epistaxis	08	1.38	01	0.17	00	00	09
Total No.	511	88.25	57	9.84	11	1.89	579

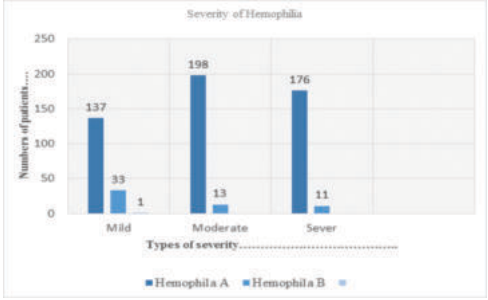


In 579cases study, most common presentation were joint bleeding 76.85% cases followed by abdominal 8% cases, muscles 7.08% cases, gum 3.45% cases, petechiae 2.24%cases, and epistaxis 1.55% cases (table 2).

In Von-Willebrand disease manifestations were mainly abdominal bleeding (8/11 cases), gum (1/11 cases) and mucocutaneous bleeding such as petechiae (1/11 cases) and epistaxis (1/11 cases). (Table 2)

Table 3. Joints Involvement In Hemophilia Patients

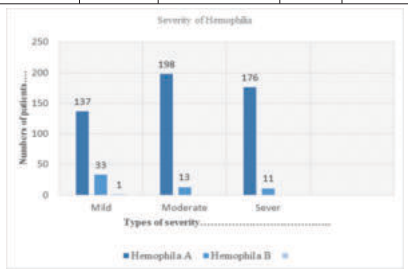
Joints	Hemophilia A	Hemophilia B	VWD	Total No.	%
Knee	203	17	00	220	54.45
Elbow	69	08	00	77	19.05
Shoulder	38	06	00	44	10.89
Others	94	10	00	104	25.74
Total No.	404	41	00	445	76.85



In 445/579 patients knee joint involvement is most common 220 (54.45%) followed by elbow 19% and shoulder 10.89% (Table 3)

Table 4. Severity of Hemophilia

Severity	Hemophilia A		Hemophilia B		Total No.
	No.	%	No.	%	
Mild 5<40%	137	28.81	33	57.89	170
Moderate 1-5%	198	38.74	13	22.80	211
Sever <1%	176	34.44	11	19.29	187
Total No.	511		57		568



In 568/579 cases (Hemophilia-A and Hemophilia-B) the percentage of mild, moderate and severe disease cases were 29.92%, 37.14% and 32.9, % respectively (table 4)

In this study Spontaneous bleed was reported in 80.13% & 9.67%% cases of hemophilia A & hemophilia B respectively.

DISCUSSION-

The age group in our study ranged from 0-42 years of age compared to less than 15 years by Uddin MM & Karim et al[11,12] & less than 18 years of age by MM Ha et al.[13] Hemarthrosis was the most frequent symptom and most frequently affected the joint was knee joint (54.45%) cases followed by elbow joint 77(19.05%) cases and shoulder joint 44(10.74%)cases, similar to Nikethan B.et al.[14] Uddin MM et al (100%) & Karim et al (82%) while MM Hazewinkel[11-13] Reported subcutaneous bleed(45%)cases as most common symptom. Additional common symptoms listed in decreasing order of frequency were joint bleeding-76.85%, cases followed by abdominal bleed 8%cases, muscles bleed 7.08% cases, gum bleed 3.45% cases, petechiae 2.24%cases, and epistaxis 1.55% cases while Uddin MM[11] reported wound bleeding (52%)cases & bleeding after tooth extraction (38%)cases, Karim et al[12] reported gum bleeding (38%) cases & bleed during surgical procedure such as circumcision & tooth extraction (28%) cases other rare presentations were hematemesis, malena, epistaxis (2%)cases while mucosal bleeding (15%)cases were reported by M H Hazewinkel.[13]

Age at first presentation in our study was less then 5 years in 58.2% cases (31.5 % were less then 1 year), While Karim MA et al[12] reported 94% cases in less than 5 years age group (64% were less than 1 years). None of the studies, including our We found positive family history in 70% of cases same as by Karim et al, while M H Hazewinkel & Uddin MM reported 49% & 30% respectively.[11-13] We have found (Hemophilia-A and Hemophilia-B) the percentage of mild, moderate and severe disease in 29.92%, 37.14% & 32.9% respectively, while MM Uddin et al[12] reported 45%, 42.5% & 12.5% as mild, moderate & severe respectively. Most important laboratory diagnostic tool was increase in APTT which was found raised in 100% of our hemophilic patients, while same had been found in majority of cases by others workers. Spontaneous bleed was reported in 57.14% & 53.33% cases of hemophilia A & hemophilia B by us, compared to 100% in severe groups & 17.5% in moderate groups by Uddin MM et al.[12]

CONCLUSION

Haemophilia A was more common than haemophilia B. The knee was the most commonly affected joint, followed by the elbow, shoulder & others joints. In healthy people, bleeding after injury is obvious, but in the severe Hemophilic, bleeding can occur even without trauma. First bleeding episode in child under the age of 3 years considered as sever category of hemophilia.

Our patient's demographics were comparable to other studies conducted in India. Our findings also show a significant improvement in care delivery for haemophilia patients

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