Poster No.: PO-T-36

Databases and registries

Inherited bleeding disorders - Experience of a not-for-profit organization in Pakistan

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Introduction

❖The epidemiology of inherited bleeding disorders in Pakistan still remains unknown.
❖Fatimid Foundation, established in 1981, is the largest not-for-profit organization in Pakistan which provides free of cost treatment to patients with inherited haematological disorders.
❖There is no online database for inherited bleeding disorders in our organization.
❖An updated registry is important for planning care and clinical trials and to assess the effective use of resources.[1]
❖This study was conducted to determine baseline epidemiologic profiles of inherited bleeding disorders registered at an non - governmental organization (NGO) in Pakistan.

Figure 1: Number of patients registered in different centres of Fatimid Foundation
***Khyber Pakhtunkhwa**

Methods

This study was performed at Fatimid Foundation, Pakistan from 1st November 2015 till 1st June 2016. Sample size was 1492. House officers at these centres were given a pro forma and retrospective data was extracted from the medical record files.

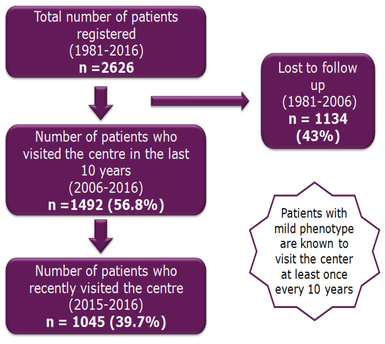

Figure 2: Number of patients registered from year 1981 -2016

Results

Centres	Diagnostic Facilities			Transfusion Transmitted Infections	Treatment Facilities			
	Screening tests (PT, APTT, Bleeding time, Platelet count)	Specific tests (Mixing studies, factor level)	Inhibitor screening*	Screening for HBV, HCV & HIV (ELISA)	Cryoprecipitate	Fresh frozen plasma	Factor concentrate (plasma derived)	Physiotherapy
Karachi	✓	✓	✓	✓	✓	✓	✓§	✓
Lahore	✓	✗	✗	✓	✓	✓	✓§	✓
Peshawar	✓	✗	✗	✓	✓	✓	✓§	✗
Quetta	✓	✗	✗	✓	✗	✓	✓§	✓
Multan	✓	✗	✗	✓	✓	✓	✓°	✓
Hyderabad	✓	✗	✗	✓	✓	✓	✓°	✗
Khairpur	✓	✗	✗	✓	✓	✓	✓°	✗
Rashidabad	✗	✗	✗	✓	✓	✓	✓°	✗

Table 1: Facilities available at each centre.
* Bethesda assay is not available at any centre § Provided free of cost in emergency ° Available on payment

Diagnosis	Males	Females	Total (%)
Haemophilia A	497	0	497 (60.8%)
Haemophilia B	77	0	77 (9.4%)
Von Willebrand Disease (VwD)	32	51	83 (10%)
Haemophilia A Carrier/ VwD	0	22	22 (2.7%)
Factor V & VIII deficiency	4	0	4 (0.5%)
Factor V deficiency	10	5	15 (1.8%)
Factor VII deficiency	2	2	4 (0.5%)
Hypofibrinogenemia	8	4	12 (1.5%)
Afibrinogenemia	2	4	6 (0.7%)
Factor II Deficiency	1	0	1(0.1%)
Factor X Deficiency	0	4	4 (0.5%)
Factor XIII Deficiency	5	2	7 (0.9%)
Glanzmann thrombasthenia	49	27	76 (37%)
Bernard Soulier Syndrome	5	5	10 (1.2%)
Total	692 (84.6%)	126 (15.4%)	**818**

Table 2: Registered patients with confirmed diagnosis (n=818)

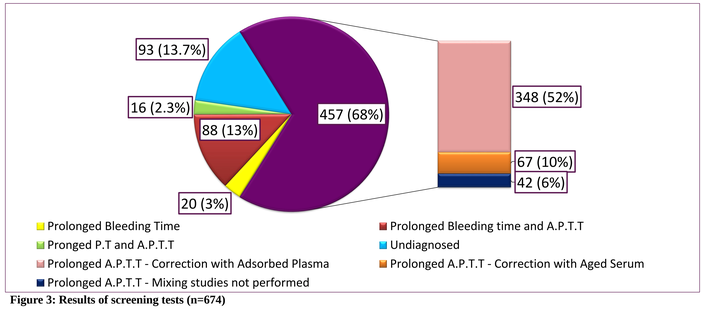

Figure 3: Results of screening tests (n=674)

Variable	n (%)
Consanguineous marriage	398/650 (61.2%)
Family history of bleeding disorder	697/1492 (46.7%)
Hypochromic Microcytic Anemia	548/1066 (51.4%)
Compliance with oral iron replacement therapy	169/548 (30.8%)

Table 3: Demographic data of inherited bleeding disorder (n=1492)

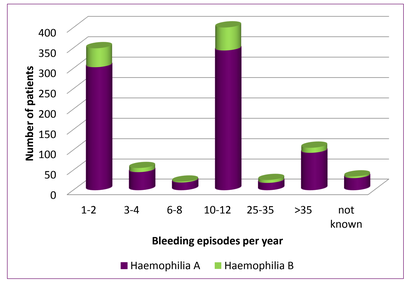

Figure 4: Number of bleeding episodes per year in haemophilia and B (n=989)

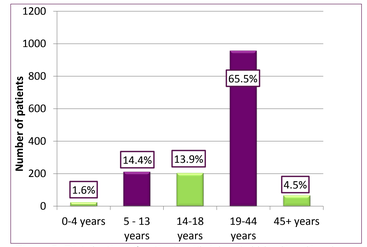

Figure 5: Distribution of patients with inherited bleeding disorder by age range (n=1461)

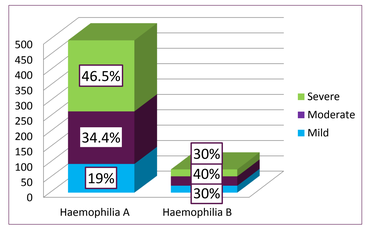

Figure 6: Severity of Haemophilia (n=574)

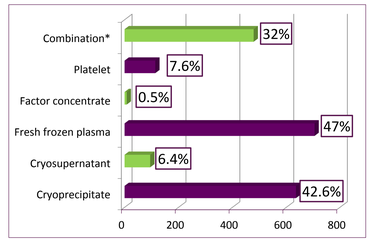

Figure 7: Treatment provided to patients (n=1492).
*** Plasma derived factor concentrate, cryoprecipitate and fresh frozen plasma**

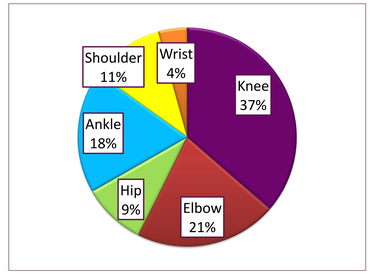

Figure 8: Haemarthrosis position (n=1032)

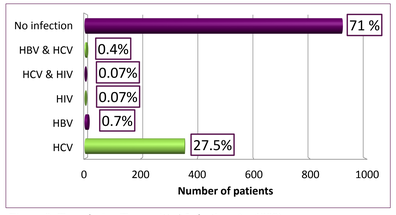

Figure 9: Transfusion Transmitted Infections (n=1289)
Immunization for hepatitis B to was provided for all patients in 2013.

Bleeding history	Diagnosis						
	Haemophilia A* (n=845)	Haemophilia B* (n=144)	VwD* (n=171)	Rare inherited coagulation disorder (n=69)	Platelet function defect (n=106)	Undiagnosed (n=157)	Total number of patients
Trauma	215	40	45	14	22	26	362
Surgery	37	7	8	6	-	6	64
Circumcision	215	40	45	14	22	26	362
Umbilical cord	4	1	3	15	-	3	26
Head Injury	50	7	11	3	4	5	80
Muscle	25	3	5	-	-	4	37
Haemarthrosis	698	122	78	38	26	70	1032
Epistaxis	207	29	95	24	71	58	484
Gums	385	65	98	38	76	79	741
Bruise	395	66	81	36	60	47	685
Genitourinary	32	3	8	3	-	4	50
Gastrointestinal	29	6	14	3	9	4	65
Menorrhagia	-	-	29	2	12	11	54

Table 4: Clinical spectrum of bleeding (n=1492)
*** Patients with presumed diagnosis based on screening tests (APTT and bleeding time) have been included with Haemophilia A, Haemophilia B and Von Willebrand disease.**

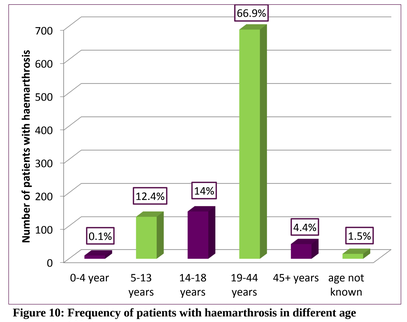

Figure 10: Frequency of patients with haemarthrosis in different age groups (n=1032)

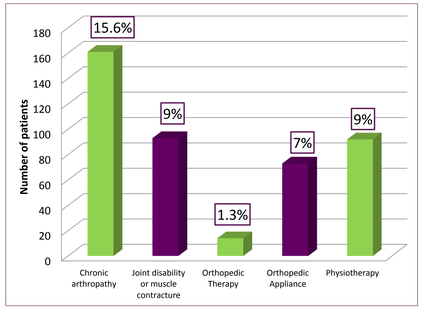

Figure 11: Complication and management of haemarthrosis (n=1032)

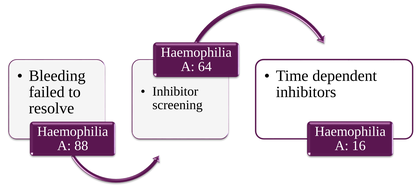

Figure 12: Results of Inhibitor screening

Conclusion

❖This is the first report of the inherited bleeding disorders from an NGO in Pakistan and the circumstances are highly similar to that of India[2].
❖Capacity building and provision of diagnostic facilities is required in most of the centres.
❖ Hemophilia patients receive on-demand therapy without any prophylaxis, contributing to high rate of musculoskeletal complications seen in adults.
❖Majority of the patients with chronic arthropathy refuse to use orthopedic appliance due to financial and social constraints.
❖ Due to the additional cost involved for orthopedic therapy, these patients are debilitated leading a poor quality of life.

Limitations:

❖Due to the manual collection of retrospective data there is a possibility of errors.
❖Around 20-30% of data was not reported either because tests were not performed or because the reports were missing from patients' record files.
❖No information was provided on mortality and factor utilization per annum.

References

[1]*Evatt BL. World Federation of Haemophilia Guide to Developing a National Patient Registry. World Federation of Hemophilia 2005.*
[2] *Ghosh K, Shetty S, Sahu D. Haemophilia care in India: innovations and integrations by various chapters of Haemophilia Federation of India (HFI). Haemophilia. 2010;16(1):61-5.*

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