Prospective And Challenges of Paediatric Hematology at King Faisal Specialist Hospital and Research Centre, Riyadh, Saudi Arabia

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Introduction:

- Benign and Pre-Malignant Hematologic Disorders constitute a major portion of the patient population seen and/or treated at the department of Paediatric Haematology / Oncology.
- According to the compiled data of last six years, on the average we are seeing 140 newly diagnosed haematology cases per year. Among these, haemophilia, thalassemia, sickle cell anaemia, bone marrow failures, VW Disease and nutritional anaemia along with thrombosis are quite common.
- This distribution clearly shows the role of our hospital as a tertiary care institution unique in providing state-of-the-art patients care for the given hematologic disorders. We are presenting demographic data of these patients at KFSH&RC for the last six years from 2005 to 2010.
- Out of a total of 1002, 200 cases of homeostasis followed by 120 new thalassemia and followed by 113 bone marrow failures, 81 with sickle cell anaemia and 59 with thrombosis.
- Most of the thalassemia patients were coming from the Eastern Province of the Kingdom of Saudi Arabia, Sickle Cell Anaemia from Jizan and Hemophilia from Riyadh.

Results:

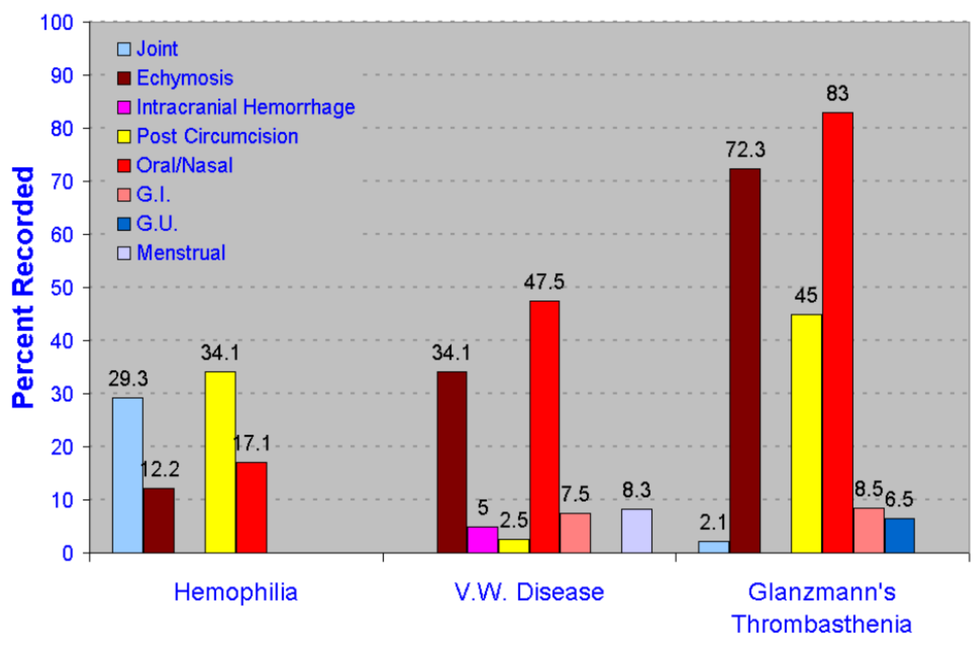
1. Demographics and Patients Characteristics

Disease Entity	Gender	Age at Dx (Median)	Family History (+)
Hemophilia	Male:97.7% (42) Female: 2.3% (1)	1.8 Years	53.5% (23)
V.W. Disease	Male:42.9% (18) Female: 57.1% (24)	5.2 Years	52.8% (19)
Glanzmann's Thrombasthenia	Male: 42.6% (20) Female: 57.4% (27)	1.8 Years	63.2% (24)
Thalassemia	Male:47.5% (57) Female: 52.5% (63)	2.3 Years	73.7% (73)
Sickle Cell Anemia	Male: 55.6% (45) Female: 44.4% (36)	4.9 Years	68% (34)

2. Serology (At Presentation)

Disease Entity	Hepatitis A (Immunity)	HBsAg (+)	Hepatitis C (Reactive)
Hemophilia	54.8%	None	6.5%
V.W. Disease	52.4%*	None**	None
Glanzmann's Thrombasthenia	-	-	-
Thalassemia	40%	1.2%	7.4%
Sickle Cell Anemia	20%	3.2%	None

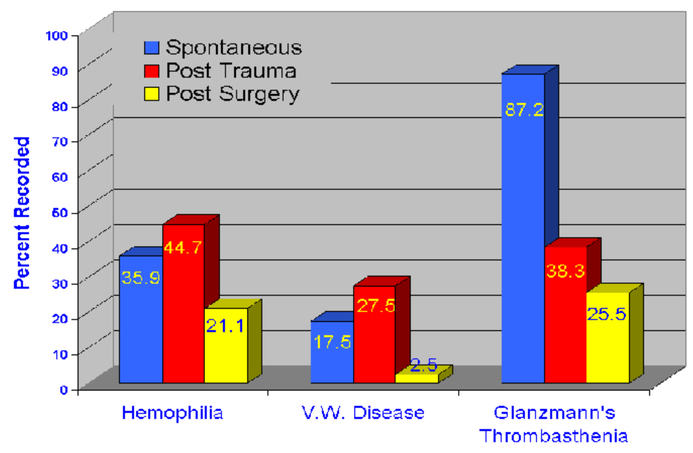
3. Presenting Complaints: Bleeding



4. Type of Disease

Disease Entity	Type		
Hemophilia	Type A: 90.7%	Type B: 9.3%	Mild: 16.7% Moderate: 16.7% Severe: 66.7%
V.W. Disease	Type 1: 54.8%	Type 2: 11.9%	Type 3: 33.3%
Glanzmann's Thrombasthenia	Type 1: 75.6%	Type 2: 4.9%	Variant: 19.5%

5. Nature of Bleeding



6. Blood Work at Presentation

Disease Entity	HgB (g/L)	PLT	PTT/aPTT
Hemophilia	121±2.7 (86-166)	437.9±28.2 (187-939)	93.02±5.7 (12.5-150) / -
V.W. Disease	117.3±3.5 (7.7-147)	355.4±12.7 (169-500)	- / 47.2±2.1 (31.7-82.4)
Glanzmann's Thrombasthenia	103.7±3.1 (40-179)	316.1±16.3 (32-563)	Prolonged: 8.7% / 8.7%

15.2% (5 of 33) patients with Hemophilia developed factor inhibitors.

* Three patient with VW Disease became Hep A Reactive during follow-up
** One patient with VW Disease became HBsAg+ during follow-up

7. Treatment Modality:

Disease Entity	Transfusions (%)	Splenectomy (%)	Bone Marrow Transplants (%)
Hemophilia	2.3% (1 of 43)	N.A	N.A
V.W. Disease	12.5% (5 of 40)	N.A	N.A
Glanzmann's Thrombasthenia	72.3% (PLT) (34 of 47)	N.A	N.A
Thalassemia	52.5% (63 of 120)	6.7% (8 of 120)	45.8% (55 of 120)
Sickle Cell Anemia	25.9% (21 of 81)	N.A	11.1% (9 of 81)

8. Complications (Druing Follow-up)

Disease Entity	Joint	Echymosis	Intracranial Hemorrhage	Post Circumcision	Nasal/Oral	G.I	G.U	Menstrual
Hemophilia	93.9% (31 of 33)	30.3% (10 of 33)	12.1% (4 of 33)	34.1% (14 of 41)	36.4% (12 of 33)	3% (1 of 33)	6.1% (2 of 33)	-
V.W. Disease	2.5% (1 of 40)	35% (14 of 40)	None	2.5% (1 of 40)	37.5% (15 of 40)	None	None	16.7% (4 of 24)
Glanzmann's Thrombasthenia	2.1% (1 of 47)	None	None	None	None	None	None	None

Conclusion:

- Role of pre-marital screening and genetic counseling is recommended.
- Prophylactic iron supplements for bleeding disorders is of vital importance.
- Early prophylactic therapy for hemophilia can avoid hemarthrosis and joint complications.
- Avoidance of traumatic delivery for bleeding disorder patients can be considered.
- Circumcision and controlling heavy menstrual flow for bleeding disorder patients should be included and implemented in the guidelines of management in the Kingdom.

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