

**PATTERN OF HAEMATOLOGICAL DISORDERS
ATTENDING HAEMATOLOGY CLINIC CHILDREN'S
HOSPITAL AIN SHAMS UNIVERSITY**

Thesis

Submitted for Partial Fulfillment of The
M.Sc. Degree in Paediatrics.

By

Suzan Ibrahim Mohamed Imam

M.B.B. Ch., (1987)

Supervised By

Prof. Dr. Ahmad Samy Khalifa

Professor of Paediatrics

Faculty of Medicine-Ain Shams University

Dr. Wagida Abdel Rahman Anwar

Assistant Professor of Public Health

Faculty of Medicine-Ain Shams University

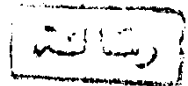
Dr. Manal Hamdy El-Sayed

Lecturer of Paediatrics

Faculty of Medicine-Ain Shams University

Faculty of Medicine
Ain Shams University

1994



بِسْمِ اللَّهِ الرَّحْمَنِ الرَّحِيمِ

"إِنَّا لَا نَضِيعُ أَجْرَ مَنْ أَحْسَنَ عَمَلًا"

صدق الله العظيم

سورة الكهف آية (٣٠)



To

My Father with

Love and Sincerity

Suzan

ACKNOWLEDGMENT

I am greatly indebted to Prof. Dr. Ahmed Samy Khalifa, Professor of Paediatrics, Ain Shams University, for his encouragement, constant guidance and honest supervision. He has been mostly generous and kindly helpful. He has spared no effort so that this thesis would see light.

I am also greatly overwhelmed by the constant help and assistance offered by Dr. Wagida Abdel Rahman Anwar, Assistant professor of Community Medicine. She has provided me with all available facilities to the statistical part of this work.

I am sincerely thankful to Dr. Manal Hamdy El-Sayed, Lecturer of Paediatrics, Ain Shams University for her continuous interest and the precious time she has given me.

I would like to convey my warmest gratitude to Dr. Azza Abdel Gawad Tantawi for her help at the beginning of this work.

I would like to express my great gratitude to Dr. Maha El-GaaFary for her help in statistical analysis.

List fo Abbreviations

G6PD-def.	Glucose-6-Phosphate Dehydrogenase deficiency
AIHA	Autoimmue Haemolytic Anaemia
ITP	Idiopathic Thrombocytopenic Purpura
M-RNA	Massanger, RNA
RES	Reticuloendothelial System
APTT	Activated Partial Thromboplastine Time
PT	Prothrombin Time
PTT	Partial Thormboplastin Time
CT	Coagulation Time
AA	Aplastic Anaemia
PRCA	Pure Red Cell Aplasia
HSP	Henoch Schenolin Purpura
AFI	Anaemia for Investigation
AHA	Acute Haemolytic Anaemia
UAHA	Unclassified Acute Haemolytic Anaemia
CHA	Chronic Haemolytic Anaemia
UCHA	Unclassified Chronic Haemolytic Aanaemia
HS	Hereditary Spherocytosis
SCA	Sickle Cell Anaemia
SCT	Sickle Cell Thalassaemia
SCD	Sickle Cell Disease
ALL	Acute Lymphocytic Leukemia
RBCs	Red Blood Cells
WBCs	White Blood Cells
ADP	Adenosine Diphosphate
VWD	Von Willebrand Disease
NCV	Mean Corpascular Volume

MCH	Mean Corpascular Haemoglobin
Hb	Haemoglobin
TIBC	Total Iron Binding Capacity
HDN	Haemolytic Disease of Newborn
HA	Haemophilia A
HB	Haemophilia B
HC	Haemophilia C
Thrombas.	Thrombasthenia
Th.	Thalassaemia
UBT	Unclassified Bleeding Tendency

List of Tables

	<i>Page</i>
Table (1) Frequency Distribution of Anaemias and Haemorrhagic Diathesis in the Haematology/Oncology Clinic Children's Hospital Ain Shams University.	82
Table (2) Frequency Distribution of Haematological Disorders.	83
Table (3) Frequency Distribution of Patients with Haematological Disorders Attending Haematology/Oncology Clinic in Children's Hospital Ain Shams University per 100'000 out Patients Clinic.	84
Table (4) Frequency Distribution of Chronic Haemolytic Anaemias.	88
Table (5) Age Distribution in Chronic Haemolytic Anaemias.	89
Table (6) Pattern of Sex Distribution in Chronic Haemolytic Anaemias.	90
Table (7) Frequency Distribution of Blood Group in Chronic Haemolytic Anaemias.	91
Table (8) Distribution of Patients of Chronic Haemolytic Anaemias Allover the Country.	92
Table (9) Pattern of Family History and Consanguinity in Anaemias.	93
Table (10) Pattern of Presentation in Thalassaemias.	94
Table (11) Pattern of Presentation in Hereditary Spheryocytosis.	95

Table (12) Pattern of Presentation in Sickle Cell anaemia.	96
Table (13) Pattern of Presentation in Sickle Cell Thalassaemia.	97
Table (14) Pattern of Liver Size in Chronic Haemolytic Anaemias (in cm).	98
Table (15) Pattern of Spleen Size in Chronic Haemolytic Anaemias (in cm).	99
Table (16) Pattern Of Reticulocytic Count in Chronic Haemolytic Anaemias.	100
Table (17) Pattern of RBCs Count in Chronic Haemolytic Anaemias (10^6).	101
Table (18) Pattern of WBCS Count in Chronic Haemolytic Anaemias (10^3).	102
Table (19) Pattern of Platelet Count in Chronic Haemolytic Anaemias (10^3).	103
Table (20) Pattern of Haemoglobin Level in Acute Haemolytic Anaemias.	104
Table (21) Pattern of Hb abnormalities in Thalassaemias and Sickle Cell Diseases.	105
Table (22) Frequency Distribution of Acute Haemolytic Anaemias.	109
Table (23) Distribution of Acute Haemolytic Anaemic Patients Allover the Country.	109

Table (24) Frequency Distribution of Blood Groups in Acute Haemolytic Anaemias.	110
Table (25) Age Distribution in Acute Haemolytic Anaemias (in month).	110
Table (26) Pattern of Sex Distribution in Acute Haemolytic Anaemia.	111
Table (27) Pattern of Presentation in G6PD-deficiency.	112
Table (28) Pattern of Presentation in Autoimmune Haemolytic Anaemia.	113
Table (29) Pattern of Presentation in Unclassified Acute Haemolytic Anaemia.	114
Table (30) Pattern of Presentation in Haemolytic Disease of Newborn.	115
Table (31) Pattern of Liver Size in Acute Haemolytic Anaemias (in cm).	116
Table (32) Pattern of Spleen Size in Acute Haemolytic Anaemias (in cm).	117
Table (33) Pattern of Reticulocytic Count in Acute Haemolytic Anaemias %.	118
Table (34) Pattern of RBCs Count in Acute Haemolytic Anaemias (10^6).	119
Table (35) Pattern of WBCs in Acute Haemolytic Anaemias (10^3).	120
Table (36) Pattern of Platelet Count in Acute Haemolytic Anaemias (10^3).	121

Table (37) Pattern of Haemoglobin Level in Acute Haemolytic Anaemias.	122
Table (38) Frequency Distribution of Other Types of anaemias.	125
Table (39) Frequency Distribution of Other Anaemias Alloner the Country.	126
Table (40) Pattern of Blood Groupes in Other Types of Anaemias.	126
Table (41) Age Distribution in Other Types of Anaemias.	127
Table (42) Pattern of Sex Distribution in Other Types of Aneamias.	128
Table (43) Pattern of Presentation in Aplastic Anaemia.	129
Table (44) Pattern of Presentation in Anaemia for Investigations.	130
Table (45) Pattern of Liver Size in Other Types of Anaemias (in cm).	131
Table (46) Pattern of Spleen Size in Other Types of Anaemias (in cm).	132
Table (47) Pattern of Reticulocytic Count in Other Types of Anaemias (%).	133
Table (48) Pattern of RBCs Count in Other Types of Anaemias (10^6).	134
Table (49) Pattern of WBCs Count in Other Types of Anaemias (10^3).	135

Table (50) Pattern of Platelet Count in Other Types of Anaemias (10^3).	136
Table (51) Pattern of Haemoglobin Level in Other Types of Anaemias (gm\dl).	137
Table (52) Frequency Distribution of Haemorrhagic Diathesis.	141
Table (53) Distribution of Patients of Haemorrhagic Diathesis Allover the Country.	142
Table (54) Pattern of Blood Groups in Haemorrhagic Diathesis.	142
Table (55) Age Distribution in Haemorrhagic Diathesis.	143
Table (56) Pattern of Sex Distribution in Haemorrhagic Disorders.	144
Table (57) Patern of Presentation in Coagulation Disorder.	145
Table (58) Pattern of Presentation in Idiopathic Throbocytopenic Purpura.	146
Table (59) Pattern of Presentation in Henoch Schenolin Purpura.	147
Table (60) Pattern of Reticulocytic Count in Haemorrhagic Diathesis (%).	148
Table (61) Pattern of RBCs Count in Haemorrhagic Diathesis (10^6).	149
Table (62) Pattern of WBCs Count in Haemorrhagic Diathesis (10^3).	150

Table (63) Pattern of Platelet Count in haemorrhagic Diathesis (10^3).	151
Table (64) Pattern of Haemoglobin Level in Haemorrhagic Diathesis (gm\dl).	152
Table (65) Frequency Distribution of Miscellaneous Group.	153

List of Figures

	<i>Page</i>
Figure (1) Frequency Distribution of Anaemias and haemorrhagic diathesis in the Haematology\Oncology Clinic, Children's Hospital Ain Shams University	82
Figure (2) Frequency Distribution of Haematological Disorders in Paediatric Haematology\Oncology Clinic, Children's Hospital, Ain Shams University	83
Figure (3) Frequency Distribution of Chronic Haemolytic Anaemias	88a
Figure (4) Pattern of Chest infection in Anaemias	97a
Figure (5) Pattern of Chest infection in Anaemias	97b
Figure (6) Frequency Distribution of Acute Haemolytic Anaemias	109a
Figure (7) Frequency Distribution of Haemorrhagic Diathesis	141a

Table of Contents

	<i>Page</i>
Introduction	1
Aim of the Work	4
Review of Literature	5
- Nutritional anaemias	11
- Sideroblastic anaemia	19
- Bone marrow failure	20
- Pure red cell aplasia	20
- A plastic anaemia	21
- Haemoglobin defects	25
- Thalassaemias	25
- Sickle cell diseases	33
- Haemolytic anaemias due to corpuscular defects	39
- Hereditary spherocytosis	39
- G6PD-deficiency	42
- Immune mediated haemolytic anaemias	49
- Haemolytic disease of new born	49
- Autoimmune haemolytic anaemia	52
- Coagulation disorders	55
- Platelet disorders	64
- Henoch Schonlein purpura	74
Subjects and Methods	75
Results	80
Discussion	154
Summary and Conclusion	176
Recommendation	179
References	180
Arabic Summary	