بِسْمِ اللّهِ الرَّحْمَنِ الرَّحِيمِ

قال الله تعالى

{ يَا أَيُّهَا النَّاسُ إِنَّا خَلَقْنَاكُمْ مِنْ ذَكَرٍ وَأُنْثَى وَجَعَلْنَاكُمْ شُعُوبًا وَقَبَائِلَ لِتَعَارَفُوا إِنَّ أَكْرَمَكُمْ عِنْدَ اللَّهِ أَتْقَاكُمْ إِنَّ اللَّهَ عَلِيمٌ خَبِيرٌ }

صدق الله العظيم

سورة الحجرات الآية 13

Dedication

To.....My beloved Parents

To .....My Brothers and Sisters

To .....sons of my Sister

To .....My Aunts and Uncles

To.....Teachers

Ibrahim

Acknowledgments

This study has been an inspiring, often exciting, sometimes challenging, and always interesting experience. Far too many people to mention individually have assisted me in many ways during my work. They all have my sincere gratitude.

I am very grateful to my supervisor Dr. Tarig El-fatih El-mesbah who has given me the chance to be one of his students; he had supported me with his keen supervision, encouragement, many fruitful discussion and regular guidance through the course of the study.

I am deeply indebted to my teachers in everywhere especially in Al Neelain University and Sudan University for Science and Technology, for their valuable experiences, advice and endless support through different ways.

Special thanks are to the staff member of the department of haematology in AL Neelain University for sharing me a parallel part of this project, their experiences in haematology as will as Research Methodology was of great value.

I am doing indebted to the staff member of AL Buluk hospital, C15 ward in Khartoum teaching hospital and staff of haematology department in national health laboratory.

List of Contents

Dedication	I
Acknowledgement	II
List of contents	III
List of abbreviations	VI
List of figures	VII
List of Tables	VIII
English Abstract	IX
Arabic Abstract	X

	Page
Chapter One Introduction and Literature Review	
1.1 Introduction	1
1.1.1 Anaemia	1
1.1.1.2 Classification of anaemia	1
1.1.1.2.1 Etiological classification	1
1.1.1.2.1.1 Impaired RBCs production	1
1.1.1.2.1.2. Increased RBCs destruction	2
1.1.1.2.1.3 Blood loss	3
1.1.1.2.2 Morphological classification	3
1.1.1.2.2.1 Hypochromic microcytic	3
1.1.1.2.2.2 Normocytic Normochromic	4
1.1.1.2.2.3 Macrocytic Anaemia	4
1.1.2 Haemoglobin	4
1.1.2.1 Genetics of Haemoglobin Synthesis	5
1.1.2.2 Normal Pattern of Haemoglobin	5
1.2.3 Haemoglobinopathies	6
1.2.3.1 Classification	7
1.2.2.2 Clinically significant variant Haemoglobins	7
1.2.2.2.1 Haemoglobin C	7
1.2.2.2.2 Haemoglobin E	8
1.2.2.2.3 Haemoglobin D	8
1.2.2.2.4 Haemoglobin O-Arab	8
1.2 Literature Review	9

1.2.1 Sickle Cell Anaemia	9
1.2.1.1 Genetics and Inheritance	9
1.2.1.2 Geographic Distribution of Sickle Mutation	10
1.2.1.3 Pathophysiology of Sickle Cell Anaemia	10
1.2.1.4 Types of Sickle Cell Anaemia	10
1.2.1.4.1 Sickle Cell Disease	10
1.2.1.4.1.1 Clinical Feature	10
1.2.1.4.1.1.1 Painful Vaso-occlusive Crises	12
1.2.1.4.1.1.2 Visceral Sequestration Crises	12
1.2.1.4.1.1.3 A plastic Crises	12
1.2.1.4.1.1.4 Haemolytic Crises	13
1.2.1.4.1.2 Possible Complication	13
1.2.1.4.1.3 Factors Affecting The Severity and Variability of Disease	13
1.2.1.4.1.4 Laboratory Diagnosis	13
1.2.1.4.2 Sickle Cell Trait	14
1.2.1.4.2.1 Clinical Feature	14
1.2.1.4.2.2 Laboratory Diagnosis	14
1.2.1.4.3 Compound Heterozygous	15
1.2.1.4.3.1 Hb SC Disease	15
1.2.1.4.3.2 Sickle cell β-thalassaemia	15
1.2.1.4.3.3 Sickle Cell Anaemia with Coexistent α-thalassaemia	15
1.2.1.4.3.4 Sickle cell HPFH	16
1.2.2 Previous Study	16
1.3 00Rationale	17
1.4 Objectives	18
Chapter two: Materials and Methods	
2.1 Study approach	19
2.2 Study design	19
2.3 Study population	19
2.4 Study area	19
2.5 Study variables	19
2.6 Samples	19
2.7 Study duration	19
2.8 Data collection method and tools	20
2.9 Laboratory procedures	20
2.10 Method of Analysis	23
2.11 Ethical Consideration	23
Chapter Three: Results	24
3.1Results	24

Chapter Four: Discussion	
4.1 Discussion	39
Chapter Five: Conclusion and Recommendation	
4.1 Conclusion	41
4.2 Recommendation	42
Chapter Six: References and Appendix	
6.1References	43
6.2 Appendixes	

Abbreviations

Hb : Haemoglobin

SCD : Sickle Cell Disease

HPFH : Hereditary Persistence of Fetal Haemoglobin

RBCs : Red Blood Cell

PCV : Packed Cell Volume

TWBCs: Total White Blood Cells

MCV : Mean Cell Volume

MCH : Mean Cell Haemoglobin

MCHC : Mean Cell Haemoglobin Concentration

HBEP : Haemoglobin Electrophoresis

List of Figures

Figure Number	Title	Page
Fig(3.1)	Showed distribution of male and female among cases	26
Fig (3.2)	Showed distribution of patient's age.	27
Fig (3.3)	showed distribution of sickle cell anaemia among tribes	28
Fig (3.4)	Showed Geographical distribution of sickle cell anaemia.	29
Fig (3.5)	Showed Residence of patients with sickle cell anaemia.	30
Fig (3.6)	Showed mean of Hb among SCD and trait.	31
Fig (3.7)	Showed mean of RBCs among SCD and trait.	32
Fig (3.8)	Showed mean of PCV among SCD and sickle trait.	33
Fig (3.9)	Showed mean of MCH among SCD and trait.	34
Fig (3.10)	Showed mean of MCHC among SCD and trait.	35
Fig (3.11)	Showed mean of MCV among SCD and trait.	36
Fig (3.12)	Showed mean of TWBCs among SCD and trait.	37
Fig (3.13)	Showed mean of platelet among SCD and trait.	38

List of Table

Table Number	Title	Page
Table (1)	Human Haemoglobins	6

Abstract

This descriptive study was carried out in different pediatric hospitals in Khartoum state during the period from November 2008 to February 2009. The study was aimed to determine the ethnical and regional distribution of sickle cell anaemia among patients referring to pediatric hospitals of Khartoum state. Complete blood count, sickling test and haemoglobin electrophoresis were performed.

A total number of 100 samples were collected from patients referring to pediatrics hospitals of Khartoum state, 81% of them with sickle cell disease and 19% with sickle cell trait based on results of haemoglobin electrophoresis. Females represent 53 of the studied patients where as male 47.

The study also aimed to determine the anaemia parameters among patients with sickle cell disease and trait and to make a comparison between sickling test and haemoglobin electrophoresis.

The results were showed that sickle cell anaemia is predominant among tribes of Western Sudan (88 % of cases) with highest frequency among Messeria tribe 31% of cases followed by Rezigat 13% and Four 11% of cases, Dbania from eastern Sudan 4% , Dinka from southern Sudan 2%.

This study showed that results of sickling test always agree with results of haemoglobin electrophoresis.

The mean values of RBCs, Hb, PCV values are decreased in sickle cell disease, in sickle cell trait mean values of Hb, PCV slightly decreased and mean RBCs are normal, while red cell indices are normal in both sickle cell disease and trait.

□□□□□□□

أجريت هذه الدراسة الوصفية في عدد من مستشفيات الأطفال بولاية الخرطوم فى الفترة ما بين نوفمبر من عام 2008 و فبراير من عام 2009 , وذلك لتحديد نسبة توزيع مرض الانيميا المنجلية بين القبائل السودانية و مناطق تواجد هذه القبائل بين المرضى المترددين على مستشفيات الأطفال بولاية الخرطوم. هدفت الدراسة أيضا إلى معرفة قياسات فقر الدم وسط المرضى. قياس الدم الكامل , اختبار الانيميا المنجلية و اختبار الفصل الكهربى للهم قلوبين تم اجراؤها.

شملت الدراسة 100 مريضا ترددوا على مستشفيات الأطفال حيث أوضحت النتائج أن 81% منهم يعانون من المرض بينما 19% منهم حاملين للمرض استنادا علي نتائج الفصل الكهربي لخضاب الدم.شكل الإناث 53من مجموع الحالات بينما شكل الرجال 43.

اثبتت الدراسة انتشارالمرض وسط قبائل غرب السودان , حيث شكلت قبيلة المسيرية اكبر نسبة لعدد المرضى 31% ثم تليت بـ قبيلتي الرزيقات 11% و الفور 9% مع وجود 6% من المرضى بشرق السودان (4%من قبيلة الدبانية و 2% من قبيلة البطاحين) و 2% من قبيلة الدينكا بجنوب السودان.

أوضحت الدراسة ايضا إن نتائج الفصل الكهربي لخضاب الدم دائما تتوافق مع نتائج اختبار الانيميا المنجلية، اضافة الى إن متوسط خضاب الدم و عدد كريات الدم الحمراء و حجم الخلايا المتراصة يكون منخفضا عن المعدل الطبيعي وسط المرضى بينما يكون طبيعيا وسط حاملي المرض.وجدت الدراسة أيضا إن معاملات كريات الدم الحمراء تكون في المعدل الطبيعي وسط المرضى و حاملى المرض ايضا .