بِسْمِ اللَّهِ الرَّحْمَنِ الرَّحِيمِ

قال الله تعالى

سَنُرِيهِمْ آيَاتِنَا فِي الآفَاقِ وَفِي أَنْفُسِهِمْ حَتَّى يَتَبَيَّنَ لَهُمْ أَنَّهُ الْجَقُّ أُولَمْ يَكْفِ بِرَبِّكَ أَنَّهُ عَلَى كُلِّ شَيْءٍ شَهِيدٌ

صدق الله العظيم

سورة فصلت الآية(53)

Dedication

To my lovely parents

To my lovely wife and my sweetly
daughters Sejoud & Malaz

They are always been the candles of my
life

My teachers, friends and colleagues

With my love

Abuzar

Acknowledgment

Primarly my thanks should be to Allah, the almighty most gracious and most merciful, who grated me the serenity, means of strength and patience to accomplish this work.

I would like to express my sincere thanks and gratitude to my supervisor **Dr. Malik Hassan Ibrahim Elfadni** for his valuable help and guidance during this study, I'm also grateful to his keen interest, patience assistance and invaluable advice.

My appreciation is extended to all the medical staff in Heglig hospital and staff of National Health Laboratory department of haematology.

My special thanks are expressed to my colleagues Nazar Abd Elhafeez, Mohammed Ali Abdalla., Ala-Eldin and Razan Mohammed. Abd. Elmotalab and to all friends who have been supported throughout this work.

Abstract

This study was analytical, descriptive and cross-sectional conducted to determine the sickle cell disease and sickle cell trait frequency among Eastern Heglig area in patients referred to Heglig hospital in Southern Kordofan state in the period between November 2008 to February 2009.

One hundred (100) patients were informed about the study and agreement for participation was obtained. A venous blood of 2.5 ml was collected in EDTA containers and investigated for sickle cell disease and sickle cell trait, a complete blood count (CBC), sickling test and haemoglobin electrophoresis were carried out.

Fully automated hematological analyzer (Sysmex Kx 21N), Electrophoresis tank and power pack were used for analysis and statistical package for social sciences (SPSS) computer program version 13 was used for data processing.

The mean age of the sickle cell disease patients was (17.2 years). The results showed that percentage of sickle cell trait and sickle cell disease were (40) and (10) respectively.

Hemoglobin level, total erythrocyte and packed cell volume of the sickle cell disease patients were (6.67g/dl), $(2.4 \times 10^6/Cumm)$ and (20.7%), respectively.

There were no significant differences in sickle cell disease and normal individuals in mean cell haemoglobin concentration (32.2g/dl), mean cell volume (86.68fl) and mean cell hemoglobin (27.98 pg) (p < 0.399, p < 0.203 and p < 0.189 respectively,

The total leukocytes (15.1X 10^3 / Cumm) p < 0.000) and platelets count (452.5X 10^3 / Cumm) p < 0.005) were significantly elevated in sickle cell disease when compared with normal individuals.

In conclusion, sickle cell anaemia is highly frequent in the studied area, which is most likely due to the consanguineous marriages.

هذه دراسة م قطعية, وصفية وتحليلية. تم إجراؤها لتحديد تردد مرض فقر الدم المنجلي وحاملي خله الكريِّه المنجليه في قبيله المسيريه المترددين علي مستشفي هجليج من منطقه شرق هجليج في الفترة ما بين نوفمبر 2008 إلي مارس 2009.

تم إعلام مائة (100) مشارك بأهداف البحث وأخذت مواف قتهم, ثم أخذت مائة عينة دم , 2.5 و

ethylene-diamine-tetra-acetic acid) مل من كل مشارك في حاويات تحتوي علي مانع التجلط (EDTA))) . تم إجراء اختبارات تعداد الدم الكامل, اختبار التمنجل و الرحلان الكهربائي للهمغلوبين.

تم استخدام جهاز (Sysmex) ر قم (Kx 21N) يعمل أوتوماتيكيا, جهاز الرحلان الكهربائي

تم استخدام جهاز (Sysmex) ر قم (Kx 21N) يعمل اوتوماتيكيا, جهاز الرحلان الكهرب للهمغلوبين و برنامج الحزم الإحصائية للعلوم الاجتماعية نسخة ر قم 13 للتحليل الإحصائي.

و قد أظهرت النتائج الإحصائية أن متوسط أعمار مرضي فقر الدم المنجلي (17.2سنة). ونسب تردد مرضي فقر الدم المنجلي وحاملي المرض 10 %, 40% على التوالي.

كما لم تُوجد فروق ذات دلالة إحصائية في متوسط تركيز الهمغلوبين للخلية (32.2 جم/100 مل p < 0.189 , متوسط همغلوبين الخلية p < 0.189 فيمتوليتر p < 0.203) ومتوسط همغلوبين الخلية (27.98).

p < 0.000 15.1 X) وتعداد الصفائح الدموية أو مجموع تعداد خلايا الدم البيضاء (p < 0.000 15.1 X) مقارنةً بأفراد أصحاء. $p < 0.005 452.5 X 10^3$ مقارنةً بأفراد أصحاء. وجُد أن تردد فقر الدم المنجلي وحاملي مرض فقر الدم المنجلي عالي في المنطقة التي تمت دراستها, وهذه الخلاصة مردها لزواج الأقارب المنتشر في تك المنطقة.

List of Tables

NO	Name of the tables	Page
1-1	Pathophysilogical classification of anemia.	5
1-2	Clinical features of sickle Hemoglobinopathies.	18
3-1	The full blood count and the age for the study	40
	population group.	
3-2	The full blood count in normal group (Hb AA), sickle	41
	cell trait (Hb AS) and sickle cell disease (Hb SS) groups.	
3-3	The full blood count and age in normal group (Hb AA)	41
	and sickle cell trait (Hb AS) group.	
3-4	The full blood count and age in normal group (Hb AA)	42
	and sickle cell disease (SS) group.	

List of Figures

NO	Name of the Figures	Page
1-1	Multifocal origin and spread of the β ^S gene.	20
3-1	The gender among the study population.	39
3-2	The hemoglobin AS, SS and AA among the study	40
	population group.	
3-3	The total erythrocytes (10^6 / μ l) in normal group (Hb AA),	42
	sickle cell trait (Hb AS) and sickle cell disease (Hb SS)	
	groups.	
3-4	The hemoglobin (g/dl) in normal group (Hb AA), sickle	43
	cell trait (Hb AS) and sickle cell disease (Hb SS) groups.	
3-5	The packed cell volume (%) in normal group (Hb AA),	43
	sickle cell trait (Hb AS) and sickle cell disease (Hb SS)	
	groups.	
3-6	The mean cell volume (fl) in normal group (Hb AA),	44
	sickle cell trait (Hb AS) and sickle cell disease (Hb SS)	
	groups.	
3-7	The mean cell hemoglobin (pg) in normal group (Hb	44
	AA), sickle cell trait (Hb AS) and sickle cell disease (Hb	
	SS) groups.	
3-8	The mean cell hemoglobin concentration (g/dl) in normal	45
	group (Hb AA), sickle cell trait (Hb AS) and sickle cell	
	disease (Hb SS) groups.	
3-9	The total leukocytes (10³/µl) in normal group (Hb AA),	45
	sickle cell trait (Hb AS) and sickle cell disease (Hb SS)	
	groups.	
3-10	The platelet count $(10^3/\mu l)$ in normal group (Hb AA),	46
	sickle cell trait (Hb AS) and sickle cell disease (Hb SS)	
	groups.	

List of abbreviations

AST	Aspartate transaminase.
CBC	Complete blood count.
CO ₂	Carbon dioxide.
Glu	Glutamic acid.
IL-2	Interleukin-2
ISCS	Irreversibly sickle cell disease.
IEF	Isoelectric focusing.
EDTA	ethylene-diamine-tetra-acetic acid
Fe **	Ferrous iron
HCT	Hematocrit
Hb	Hemoglobin
Hb S	Hemoglobin S
Hb A	Hemoglobin A
Hb A ₂	Hemoglobin A ₂
Hb E	Hemoglobin E
Hb F	Hemoglobin F
HPFH	Hereditary persistence of fetal hemoglobin
HPLC	high performance liquid chromatography
LCD	Liquid crystals display.
MCV	Mean cell volume
MCH	Mean cell haemoglobin
MCHC	Mean cell hemoglobin concentration
O_2	Oxygen
PCV	Packed cell volume
PLT	Platelet
PF	Plasmodium falciparum
RBC	Red blood cell.
RDW	Red blood cell distribution width
SCA	Sickle cell anemia.
SCD	Sickle cell disease
SPSS	Statistical package for social sciences
TNF-α	Tumor necrosis factor-alpha
TWBCs C	Total white blood cells count.
TRBCs C	Total red blood cells count.
Val	Valine
WBC	White blood cell.

List of contents

NO	CONTENTS	Page
آڍة		I
Dedication.		
Acknowledgment.		III
Abstract in English.		IV
Abstract in Arabic.		V
List of tables.		VI
List of figures.		VII
List of abbreviations.		VIII
List of	contents.	IX
	CHAPTER ONE	
	Introduction and literature review	
1-1	Introduction.	1
1-2	Clinical Features of anemia.	3
1-3	Classifications of anemias	3
1-4	Physiological adaptations in anemia	5
1-5	Normal hemoglobin	7
1-6	Hemoglobinopathies	8
1-7	Nomenclatures	9
1-8	The sickle Hemoglobinopathies	10
1-9	History	11
1-10	Genetics and inheritance of Hemoglobinopathies	13
1-11	Pathophysiology	16
1-12	Clinical features	17
1-13	Prevalence and geographic distribution	18
1-14	Sickle Cell Crises	21
1-15	Other Complications of Sickle Cell Disease	22
1-16	Variant sickle cell syndromes	25
1-17	Other sickling syndromes	27
1-18	Laboratory Testing	28
1-19	Rationale.	30
	Objective.	31
	CHAPTER TWO	
	Methodology	
2-1	Study design.	32
2-2	Study area.	32
2-3	Study population.	32
2-3-1	sampling.	32
2-3-2	inclusion criteria.	32

2-3-3	exclusion criteria.	32			
2-3-4	Sample size.	32			
2-4	Tools of data collection.	32			
2-5	Data analysis.	32			
2-6	Ethical consideration.	33			
2-7	Methodology.	33			
2-7-1	Method of sample collection.	33			
2-7-2	Method of automated analyzer system (Complete	34			
	haemogram).				
2-7-3	Method of Preparation and staining of blood films:	35			
2-7-4	Sickling Test.	36			
2-7-5	Electrophoresis method.	36			
	CHAPTER THREE				
	The results				
3	Results.	39			
CHAPTER FOUR					
Discussion, Conclusion and Recommendation					
4-1					
4-1	Discussion.	47			
4-1	Discussion. Conclusion.	47 51			
4-2	Conclusion.	51			
4-2	Conclusion. Recommendations.	51			
4-2	Conclusion. Recommendations. CHAPTER FIVE References	51			