

□□□□

بسم الله الرحمن الرحيم

قال الله تعالى

سورة القيامة

أَلَمْ يَكُنْ نُطْفَةً مِّن مَّنِيِّ يَمْنَىٰ ﴿٣٧﴾ ثُمَّ كَانَ عَلَقَةً فَخَلَقَ فَسَوَّىٰ ﴿٣٨﴾ فَجَعَلَ مِنْهُ
الزَّوْجَيْنِ الذَّكَرَ وَالْأُنثَىٰ ﴿٣٩﴾ أَلَيْسَ ذَلِكَ بِقَدْرِ عَلَىٰ أَنْ يُحْيِيَ الْمَوْتَىٰ ﴿٤٠﴾

صدق الله العظيم

Dedication

To

The Soul of my Father, My lovely Mother, My Dear Husband, Sisters,

Brothers, Colleagues, & friends who have always been the candles until I

complete this work.

Acknowledgment

Primary my prays and 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 deeply obliged to my supervisor **Dr. Mahmoud Mohamed Elgari** 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 laboratory staff in Omdurman Pediatric hospital department of hematology and staff of Biochemistry department college of Medicine Khartoum University.

My special thanks are expressed to hematology staff in Sudan University of Science & Technology, to all my colleagues, & friends who have been very supportive throughout this work.

Abstract

This analytical, descriptive and cross-sectional study aim to evaluate the hematological parameters of sickle cell disease and sickle cell trait gene patients compared to control individuals admitted to Omdurman Pediatric hospital of Emergency in Khartoum state in the period between April 2011 to July 2011.

One hundred (100) patients, and (50) controls were informed their parents about consent for participation was obtained. A venous blood of 2.5ml was collected in ethylene-diamine-tetra-acetic acid (EDTA) containers and investigated for sickle cell disease and sickle cell trait, a complete blood count (CBC) [semi automated hematological analyzer (Sysmex kx21N)] was used, also blood film, correction of WBCs, reticulocyte count, and hemoglobin electrophoresis were carried out, and statistical package for social sciences (SPSS) computer program version 16 was used for data processing.

The percentage of HbAA according to the gender (70%) in the males and (30%) in the females, HbAS (20%) of the males (16%) of the females. HbSS (33%) and (28%) in the males and females respectively. HbSF (2%) of the males and (1%) of the females.

The percentage of Hb type according to the age (30%) of HbAA were <5 years, (36%) were between 5 and 10 years and (34%) were >10 years. For HbAS (11%) were <5 years, (15%) were between 5 and 10 years and (10%) were > 10 years. 27% of HbSS were <5 years, 25% were between 5 and 10 years and 9% were >10 years. All the frequency of Hb SF (3%) were found to be <5 year.

The means of Hb, RBCs & PCV were significantly increase in Hb AA than in Hb SS while the means of platelets count, reticulocytes count, & total leukocyte counts were significantly decrease in Hb AA than in Hb AS.

There were found significant increase in Hb AS than Hb AA in the neutrophil count with the p value (< 0.05) and mixed with the p value(<0.05). Other parameters show no difference between Hb AS and Hb AA.

This results showed that means of Hb, RBCs & PCV were significantly decrease in Hb SF than in Hb AA while the means of PLTs count, absolute neutrophil and mixed were significantly increase in Hb SF than in Hb AA.

The means of Hb, RBCs & PCV were significantly decrease in Hb SS than in Hb AS while the means of platelets count, reticulocytes count, & total leukocyte count were significantly increase in Hb SS than in Hb AS.

الخلاصة

هذه دراسة تحليلية وصفية مقطعية لتقويم المعايير الدموية لدى مرضي فقر الدم المنجلي وحاملي الجين م مقارنة بأفراد أصحاء محالين إلي مستشفى الأطفال بولاية الخرطوم بين ابريل 2011-يوليو 2011

استعمل (EDTA) جمعت 2.5مل عينة دم وريدي في حاويات تحتوي علي مانع التجلط ايضا مسحة الدم, وتصحيح الخلايا, (Sysmex KX-21) الجهاز الشبه آلي للتحليل الدموي

البيضاء، الخلايا الشبكية وايضا اختبار تمرحل خضاب الدم والتحليل الاحصائي بواسطة برنامج الحزم الإحصائية 16 للحصول علي البيانات

للجنس (70%) ذكور و (30%) إناث . ونسبة نوع خضاب (AA) نسبة نوع خضاب الدم (SF) . ذكور و (28%) إناث (33%) (SS) . للجنس (20%) ذكور و (16%) إناث (AS) الدم . ذكور و (1%) إناث (2%)

كانت (AA) نسبة خضاب الدم لدي الفئات العمرية كانت (30%) من نوع خضاب الدم أعمارهم اقل من خمسة أعوام , (36%) من خمسة إلى عشرة أعوام , (34%) كانت أكثر كانت (11%) وكانوا اقل من خمسة (AS) من عشرة اعوام . خضاب الدم من النوع أعوام , (15%) من خمسة الي عشرة أعوام , (10%) كانوا أكثر من عشرة اعوام . (27%) كانت أعمارهم اقل من خمسة أعوام , (25%) أعمارهم من (SS) من نوع خضاب الدم خمسة الي عشرة , (9%) كانوا أكثر من عشرة أعوام . و كانت نسبة أعمار خضاب الدم . وجد أن أعمارهم اقل من خمسة اعوام (3%) (SF)

متوسط خضاب الدم , عدد الكريات الحمراء , وحجم تكس الخلايا كان يزيد بزيادة ذات دلالة بينما كان عدد الصفائح الدموية و (SS) عن النوع (AA) احصائية في خضاب الدم النوع الخلايا الشبكية والعدد الكلي للخلايا البيضاء يقل بدلاله احصائيه في خضاب الدم من توجد زيادة ذات دلالة إحصائية بين نوعي خضاب الدم . (SS) عن النوع (AA) النوع في متوسط الخلايا المتعادلة باحتماليه أقل من (0.05) وأيضا متوسط (AS) , (AA) (الخلايا الخليط (الخلايا الوحيدة, الخلايا القاعدية, الخلايا الحمضية

هذه النتائج أوجدت أن متوسط تركيز خضاب الدم, التعداد الكامل للخلايا الحمراء وحجم أقل بدلالة إحصائية واحتمالية (0.05) عن نوع (SF) تكس الخلايا في خضاب الدم النوع بينما زاد متوسط التعداد الكامل للصفائح الدموية, التعداد الكامل للخلايا (AA) خضاب الدم بينما لا يوجد فرق في متوسط التعداد , (AA) البيضاء المتعادلة والخلايا الخليط عن النوع (AA) , (SF) الكلى للخلايا الحمراء والخلايا اللمفاوية في نوعي خضاب الدم

متوسط كل من تركيز خضاب الدم, التعداد الكامل للخلايا الحمراء و حجم تكس الخلايا بينما (AS) يقل بدلالة إحصائية واحتمالية أقل من (0.05) عن النوع (SS) لخضاب الدم يزيد متوسط كل من تعداد الصفائح الدموية ,متوسط التعداد الكلى للخلايا الشبكية والتعداد الكلى للخلايا البيضاء و يزيد بدلالة إحصائية و احتمالية أقل من (0.05) في خضاب الدم (AS) عن النوع (SS) النوع

Contents

Contents	Page
الآية	I
Dedication	II
Acknowledgment	III
Abstract in English.	IV
Abstract in Arabic.	VI

List of contents.	VIII
List of tables.	XI
List of abbreviations.	XII
CHAPTER ONE	
INTRODUCTION AND LITERATURE REVIEW	
1.1Introduction	1
1.2Epidemiology and Risk Factors	3
1.3Genetic	4
1.4History of Sickle Cell Disease	5
1.5 Causes	6
1.6Classification of sickle disease	7
1.6.1Homozygous HbSS (sickle cell anaemia)	7
1.6.2Heterozygous HbAS (sickle cell trait)	7
1.6.3Other forms of SCD	7
1.6.3.1Sickle cell HbSC	7
1.6.3.2Sickle cell β -thalassemia	8
1.6.3.3Sickle cell with hereditary Persistence of F haemoglobin(SPFH)	8
1.7Pathophysiology	9
1.8Factors That Influence the Severity of Sickle Cell Disease	9
1.9Laboratory Value	13
1.10Clinical Manifestations and Complications of Sickle Hemoglobin	14
1.11Sickle cell crisis	16
1.11.1Painful vaso-occlusive crises	16
1.11.2Visceral sequestration crises	16
1.11.3Aplastic crises	17
1.11.4Hemolytic crises	17
1.11.5 Other clinical feature:-	17
1.12Other clinical feature	17
1.12 Normal human haemoglobin	18
1.12.1 Haemoglobin A (Hb-A)	18
1.12.2Haemoglobin A2 (Hb- A2)	18
1.12.3Haemoglobin F (Hb-F)	18
1.12.4Embryonic hemoglobin	19
1.12.5abnormal hemoglobin's	20
1.13Diagnosis of Sickle Hemoglobin	20
1.14 Routine Health Maintenance	22
1.15Objectives	24
1.16Rationale	25
CHAPTER TWO	
Materials & Methods	
2.1Study design.	26

2.2Study area.	26
2.3Study population.	26
2.4Sampling.	26
2.5Inclusion criteria.	26
2.6Exclusion criteria.	26
2.7Sample size.	26
2.8Tools of data collection.	27
2.9Data analysis.	27
2.10Ethical consideration	27
2.11Methodology.	27
2.11.1 Method of sample collection.	27
2.11.2Method of automated analyzer system (Complete hemogram).	28
2.11.3Method of Preparation and staining of blood films:	29
2.11.4 Reticulocyte Count	30
2.11.5Sickling Test.	32
2.11.6Electrophoresis method	33
CHAPTER THREE The results	
3. Results.	35
CHAPTER FOUR Discussion, Conclusion and Recommendations	
4.1 Discussion.	43
4.2 Conclusion.	46
4.3 Recommendations.	47
CHAPTER FIVE References	
5.1 References.	48
5.2 Appendices.	53

List of Tables

Table No	Title	Page No
(1.1)	Sickle diseases	8
(1.2)	The normal hemoglobin's	19
(1.3)	The common abnormal hemoglobin's	20
(2.1)	Reticulocyte Production Index Correction Factors	31
(3.1)	Types of Hb in study group according to the gender	35
(3.2)	Types of Hb in control group according to the gender	35
(3.3)	Frequency of age group among study group	36
(3.4)	Frequency of age group among control group	36
(3.5)	Comparison between test & control group	37
(3.6)	Comparison between Hb AA & Hb SS	38
(3.7)	Comparison between Hb AA and Hb AS	39
(3.8)	Comparison between Hb AA and Hb SF	40
(3.9)	Comparison between Hb AS and Hb SS	41
(3.10)	Results of hematological parameters in different Hb type	42

List of Abbreviations

- CBC : Complete Blood Count.
- EDTA : Ethylene Di amine Tetra acetic Acid.
- Fe²⁺ : ferrous Ion.
- fL : Fimto Littre.
- G-6-PD: glucose-6-phosphate dehydrogenase.
- Glu-gln : Glutamine-Guanine.
- Hb S : Sickle Hemoglobin .
- Hb SC : Hemoglobin S & C.
- HbA : Adult Hemoglobin.
- HbF : Fetal Hemoglobin.
- Lys : Lysine.
- mA : milli Ampire.
- MCH : Mean corpuscular hemoglobin .
- MCHC : Mean corpuscular hemoglobin concentration .
- MCV : Mean corpuscular volume .
- RBCs: Red Blood Cells.
- RDW : Red Diameter Width.

- SCD : Sick Cell Disease
- SCD-HbAS : Sick Cell Disease Hemoglobin AS.
- SCD-HbSC : Sick Cell Disease Hemoglobin SC.
- SCD-HbSS : Sick Cell Disease Hemoglobin SS.
- SPFH : Sick cell with hereditary Persistence of F haemoglobin.
- SPSS : statistical package for social sciences.
- TEB : Tris Ethylene Di amine Tetra Acetic Acid Borate .
- [USA](#) : Unite State of America.
- V : Volt.
- Val : Valine.
- WBC : White Blood Cell.