

بِسْمِ اللَّهِ الرَّحْمَنِ الرَّحِيمِ

فَقَالَ تَعَالَى

وَيَسْأَلُونَكَ عَنِ الرُّوحِ فُلِّ الرُّوحِ مِنْ أَمْرِ رَبِّيِّ وَمَا} {أَوْتَيْتُمْ مِنَ الْعِلْمِ إِلَّا قَلِيلًا

(الإسراء : ٨٥)

Dedication

To those who are bright as the moon, their heats shine
With light of million heavily stars and who are always
encouraging in many pursuit

My parents

*

*

To my lovely brothers & sisters

To my friends in study and working life

To supervisor who guidance made such progression in my
life

For All patients in the hope of treating the sickle cell
haemoglobinopathies

Acknowledgment

Praise to Allah the almighty who gave us the health and strength.

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Abstract

This descriptive case control study was conducted to determine the types, patterns and level by percent (%) of the haemoglobins among Sudanese patients with sickle cell haemoglobinopathies through the period of March 2012 up to May 2012, using Capillary Electrophoresis technology. The total number of study group was one hundred, seventy patients group and thirty as control group. 51.4% of patients group were female while 48.6% were male. The study reveals the distribution rate of sickling cell haemoglobinopathies among all age groups. The study results shows that the frequency of AS patterns were 52.9%, AS/C patterns were 1.4%, S/BThalassemia patterns were 11.4%, SC patterns were 1.4%, SD patterns were 4.3% and SS patterns were 28.6%. However, all patients with sickle cell haemoglobinopathies might have the abnormal form of haemoglobin (HbS) even in small amount. Hence, the amount of HbS (Mean) show 39.23% of AS pattern, 42.8% of ASC pattern, 47.5% of SC pattern, 11.64% of S/ β Thalassemia, 56.3% of SD pattern and 84.1% of SS pattern. The study also reveals that the mean of haemoglobin A pattern in patients group were statistically significant lower than means of control group (P value < 0.05).

The study also explain the rate effect of HbF on the type of treatment, if the mean was 13.62 % and it is statistically significant higher than means of control group (P. value > 0.05).

In sickle cell haemoglobinopathies (with exception S/βThalassemia) non significant differences of haemoglobin A2 in comparison with control group (P. value > 0.05), while Hb F and Hb S show significant elevation respectively in comparison with control group (P. value < 0.05).

ملخص البحث

أجريت هذه الدراسة التحليلية الوصفية لتحديد أنواع وأنماط وكمية خضاب الدم بالنسبة المئوية عند المرضى السودانيين المصابين بخضاب الدم المنجلی المعتل في الفترة ما بين مارس 2012 حتى مايو 2012 باستخدام تقنية الرحلان الشعيري الكهربائي. مجموع عدد الحالات في الدراسة مائة ، سبعون مجموعة المصابين وثلاثون يمثلون المجموعة الضابطة. مثلت الإناث نسبة 51,4 % من المجموعة المصابة بينما مثل الذكور 48.6 %. كشفت الدراسة ان معدل الاصابة بخضاب الدم المنجلی المعتل يشمل جميع الفئات العمرية.

أظهرت نتائج الدراسة ان تكرار نمط الخضاب ذو الرحلان الشعيري hT- β\ S النمط ، (1.4%) C\SA والنمط ، (52.9%) SA الكهربائي SS و النمط (4.3%) DS النمط ، (1.4%) CS والنمط ، (11.4%) كما أظهرت ان جميع المرضى المصابين بخضاب الدم . (28.6%) وان كان بكمية S المنجلی المعتل لديهم الخضاب ذو الرحلان الكهربائي في المجموعة ذات النمط S بسيطة، وبالتالي كان متوسط النمط SA في مجموعة النمط S 39.23% ، 42.8% ، C\SA في مجموعة النمط 47.5% ،

في 56.3% hT- β\S ، 11.64% CS ، النمط SS. و 84.1% في مجموعة النمط DS مجموعة النمط. كذلك أوضحت الدراسة انخفاض معدل متوسط الخضاب ذو الرحلان في المجموعة المصابة مقارنة بالمتوسط المتحصل عليه من A الكهربائي المجموعة الصابطة ، ووجد أنه أقل بفرق احصائي ذو دالة معنوية (القيمة المعنوية < 0.05).

في F كذلك أوضحت الدراسة أثر زيادة الخضاب ذو الرحلان الكهربائي المجموعة المصابة والذي اذا كان متوسطه 13,6% بالمقارنة مع الرحلان الكهربائي عند المجموعة الصابطة والفرق ذو دالة احصائية معنوية (القيمة المعنوية < 0.05).

في مرضى فقر الدم المنجلی المتجانس (باستثناء فقر الدم التلاسيمي) و A وجد أنه لا اختلاف بين متوسط الرحلان الكهربائي لخضاب الدم نوع 2 متوسطه في المجموعة الصابطة ، بينما يوجد فرق احصائي بين متوسط حيث كانا مرتفعان في S و النوع F الرحلان الكهربائي لخضاب الدم نوع (مرضى فقر الدم المنجلی المعتل (القيمة المعنوية < 0.05).

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List of Abbreviations

δ-ALA	δ-aminolevulinic acid
BCSH	British Committee for Standards in Haematology
BMT	Bone Marrow Transplantation
CE	Capillary Electrophoresis
CZE	Capillary Zone Electrophoresis
CO ₂	Carbon dioxide
CA	Carbonic Anhydrase
CoA	Coenzyme A
Cl ⁻	Chloride ions
CT	Computerized Tomography
DNA	Deoxyneuclic Acid
2,3-DPG	2,3Dibhosphoglycerate
ED	Emergency Department
EDTA	Ethylenediaminetetraacetic acid
EMS	Emergency Medical Services
EOF	ElectroosmoticFlow
FEP	Free Erythrocyte Protoporphyrin
FDA	Food and Drug Administration
Hb	Haemoglobin
HPLC	High-Performance Liquid Chromatography
HPCEC	High-Performance Cation-Exchange Chromatography
HLA	Human Leukocyte Antigen

H^+	Hydrogen
HU	Hydroxyurea
HPFH	Hereditary Persistence of Fetal Haemoglobin
IgG	Immunoglobulin G
IRDS	Infant Respiratory Distress Syndrome
ISCs	Irreversibly Sickled Cells
IEF	Iso-electro Focusing
Kg	Kilogram
MCH	Mean Concentration of Haemoglobin
MCHC	Mean Cell Haemoglobin Content
MCV	Mean Cell Volume
MECC	Micellar Electrokinetic Capillary Chromatography
mg	milligram
MRI	Magnetic Resonance Imaging
mRNA	Massenger Ripe nucleic acid
NO	Nitric Oxide
NRBCs	Nucleated Red Cells
O_2	Oxygen
pH	Power of Hydrogen
RES	Reticuloendothelial System
SC*	Sickle cell Hb C disease with Hb A
SC	Sickle cell Hb C disease
SD	Sickle cell Hb D disease
SCD	Sickle Cell Disease
US	United State
VCAM-1	Vascular Cell Adhesion Molecule-1