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Full Length Research Paper

Frequency of the sickle cell disease and sickle cell trait in Heglig Area- Sudan

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Sickle cell disease is a genetic, hereditary and chronic disease that affects the health of its carriers and might impair their health-related quality of life. The aim of the current study was to determine the sickle cell trait frequency in Sudanese patient living in Heglig area in Western of Southern Kordofan state from November 2008 to February 2009. An analytical, descriptive and cross-sectional study conducted for one hundred participants who had confirmed patient diagnosed as (Hb S disease) homozygosis patient,2-5 mL of venous blood was collected for the measurements of complete blood count, sickling test and Hb electrophoresis. Demographic data and family history were collected in a pre designed questionnaire with written consent all participants. In this study the frequency of sickle cell trait and sickle cell disease were (52%) and (14%) respectively and 34% were normal. The sickling test showed that 71 % of the study population were negative sickling test, the remaining 29 % were positive. The total erythrocytes was significantly decreased in sickle cell disease (p< 0.000) compared with normal and sickle cell trait, also the hemoglobin concentration and packed cell volume were significantly lower than that of normal individuals and sickle cell trait patients. The frequencies of sickle cell trait was higher among the participants and patients of sickle disease showed lower values of red blood cells parameters , but higher values of white blood cells and platelets compared to haemoglobin phenotype AA control participants.

Key words: Sickle cell disease, Frequencies, Haematological values, Heglig , Sudan.

INTRODUCTION

Sickle cell disease is a major public health disease that has great impact on both individuals and society. Sickle cell disease is also associated with serious morbidity and mortality under unusual circumstances Sears DA,(1978&1994); Biedrzycki OJ& Sheaff M. (2010).Sickle cell trait (SCT) occurs at about 8% in American Blacks and from 20- 50% among some African tribes Sickle cell disease (SCD) is a more common and severe disease in Africa. Nigeria the most populous black nation in Africa has the largest number of sickle cell anaemia (SCA) patients in the world Ambe JP et.al.,(2012).In Sudan the high prevalence were reported from Western and Southern regions, where the gene frequency of this disease is quite prevalent and high rate was detected among the Baggara tribe group that includes Hawazma and Meseria Hoffbrand AV,et.al.,(2006).Most of the detected cases of sickle cell anaemia,came to the hospital with clinical symptoms unfortunately, the dangerous groups are sickle trait whom act as an instrumental for the propagation of this disease among the society through consanguineous marriages that is traditionally favoured in Sudan especially in the Western Sudan. The frequency of sickle cell trait has not been studied satisfactorily in Sudan especially in the western part where the gene frequency of sickle cell disease is quite prevalent This Study aimed to detect the sickle cell trait patients by using modern techniques in

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Figure 1. The Frequency of sickle cell trait AS,sickle cell diseases SS as compared with normal population AA.

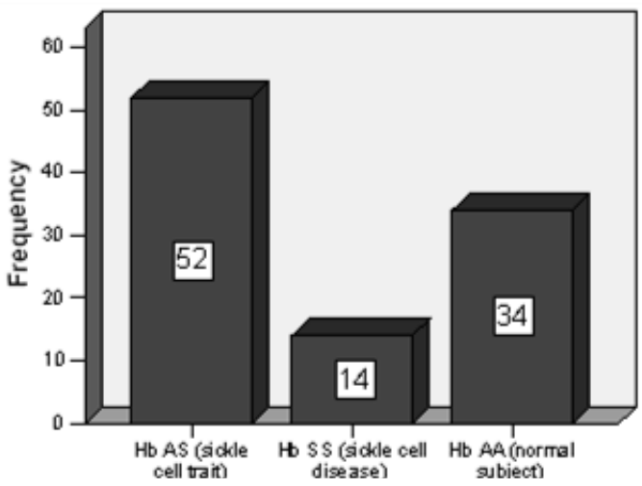


Table 1. The Age and Frequency of Participants.

The age	Frequency (%)
Less than 5 years	21
6-16 years	30
16-27 years	20
27-48 years	24
48-68 years	5
Total	100

Heglig area in Western of Southern Kordofan state to control the disease, set people aware of the genetic consequences of inbreeding, and improve behaviour and attitude towards consanguineous marriages.

MATERIALS AND METHODS

Study design

An analytical, descriptive and cross-sectional study conducted to determine the sickle cell trait frequency in Sudanese patient living in Heglig area in Western of Sothern Kordofan state during the period of November 2008 to February 2009.

Study population

All families which have at least one individual diagnosed of sickle cell disease and belong to Meseria Tibe who agreed to participate in this study were investigated for sickle cell disease and sickle cell trait. Inclusion criteria were all families who have confirmed patient diagnosed as (Hb S disease) homozygosis patient. The study sample size was hundred (100) patients.

Blood Sample Collection

A blood sample of 2-5 mL was collected from all participants into EDTA container for the measurements of complete blood count using Sysmex Kx 21N automated hematological analyzer, sickling test and Hb electrophoresis following laboratory routine procedures as described in Barbara J Bain,(2006);Lewis M,et.al.,(2006). Demographic data and family history were collected in a pre designed questionnaire.

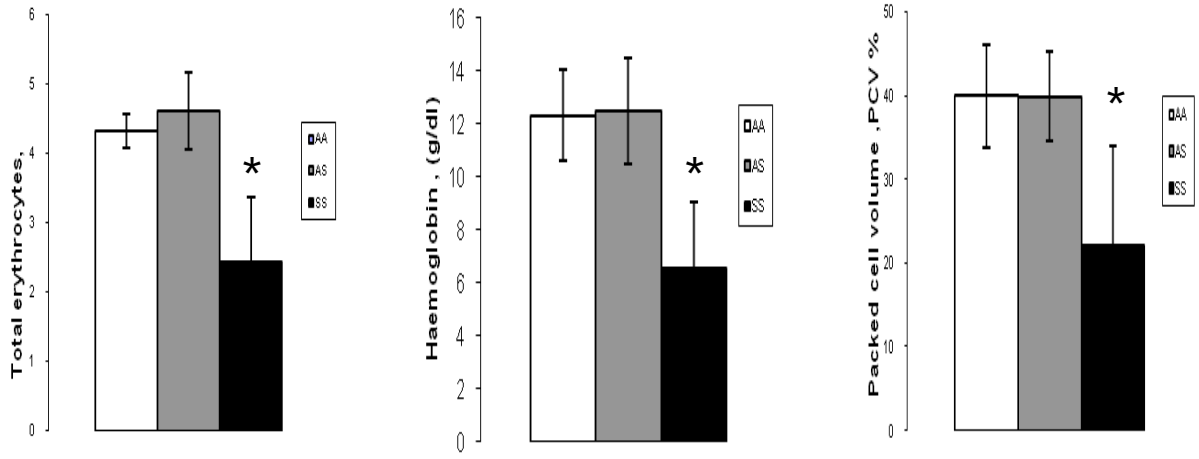
Data analysis

Data was analyzed, using statistical package for social

Table 2. The mean and SD for complete blood count (CBC).

The parameters	Mean and SD
Total erythrocytes (10^6/ µl)	4.21±1.02
Haemoglobin (g/dl)	11.61±2.80
Packed cell volume (PCV) or Haematocrit (Hct) %	37.39±8.12
Mean cell haemoglobin concentration (MCHC) g/dl	30.91±1.63
Mean cell volume (MCV) fl	88.23±7.71
Mean cell haemoglobin (MCH) pg	27.31±3.07
The total leukocytes (10^3/µl)	8.80±7.23
Platelet count (10^3/µl)	243.60±102.15

Figure 2. Total Erythrocytes (10^6/ µl), Haemoglobin g/dl) and Paced Cell Volume (PCV)% for normal group (Hb AA), sickle cell trait (Hb AS) and sickle cell disease (Hb SS) groups.



sciences (SPSS) computer program version 13 for mean, standard deviation and frequencies.

RESULTS

All the study participants were Meseria from Heglig area in Southern West of Kordofan, their mean age was 18 year ,71% were aged less than 30 years ,ratio of male to female was 44:54, (Table -1). The frequency of sickle cell trait (Hb AS)and sickle cell disease(Hb SS) were 52% and 14% respectively with 34% normal Hb AA as in (Figure.1).

The total erythrocytes was significantly decreased in sickle cell disease ($p < 0.000$) when compared with normal and sickle cell trait, also the haemoglobin concentration and packed cell volume were significantly lower than that of normal individuals and sickle cell trait patients.

There were no significantly differences in sickle cell trait and normal individuals in mean cell haemoglobin concentration, mean cell volume and mean cell haemoglobin.

The total leukocytes and platelet counts was significantly elevated in sickle cell disease when compared with normal individuals.

DISCUSSION

The sickle cell anemias are suggested to play an important role in the society of Heglig area where it affe-

Figure 3. Mean Cell Haemoglobin Concentration (MCHC),g/dl ,Mean Cell Volume (MCV) fl ,Mean Cell Haemoglobin (MCH) pg for normal group (Hb AA), sickle cell trait (Hb AS) and

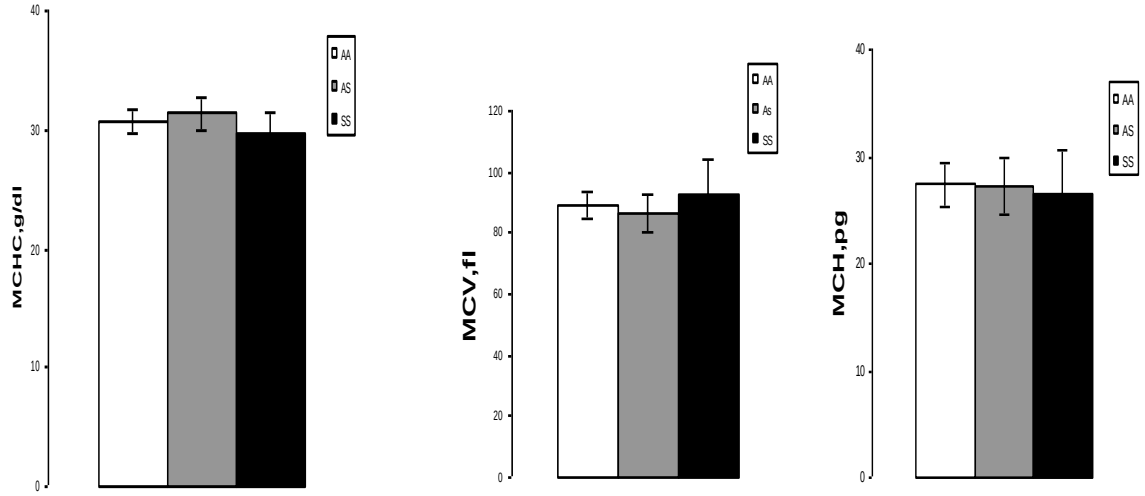
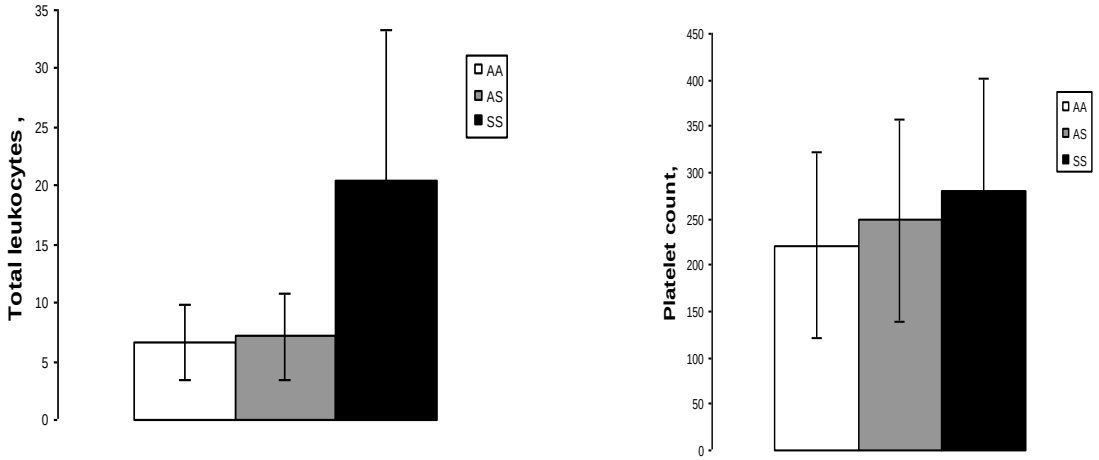


Figure 4. The Total Leukocytes (10^3/µl) and Platelet Count (10^3/µl) for normal group (Hb AA), sickle cell trait (Hb AS) and sickle cell disease (Hb SS) groups.



cts the public health. This study was conducted to investigate the sickle cell disease and sickle cell trait among Meseria tribe in Heglig area in Southern West of Kordofan.One hundred individuals of Meseria were investigated for, sickling test, full blood count and haemoglobin electrophoresis.

The mean age of the sickle cell disease patients was significantly lower than that of normal individuals (haemoglobin AA) and sickle cell trait patients ($p < 0.008$), there was suggestion of a direct relationship between the young age and the sickle cell disease. The total erythrocytes, haemoglobin concentration and packed cell volume were significantly decreased in sickle cell disease ($p < 0.000$) when compared with normal and sickle cell trait, which indicated to anaemia in sickle cell disease patients, this results agreed with previous study which reported that anaemia is significantly more often in patients with sickle cell disease compared with sickle cell trait at all ages 3 months and older,Losek JD,et.al.,(1992).

There were no significantly differences in sickle cell trait and normal individuals in mean cell haemoglobin concentration, mean cell volume and mean cell haemoglobin. This indicated no morphological differences between sickle cell trait (AS) and normal (AA) individuals. Similar results was obtained by John et al 2003, who

Table 3. The mean and SD for Complete Blood Count(CBC) for normal group (Hb AA), sickle cell trait (Hb AS) and sickle cell disease (Hb SS) groups.

The parameters	Mean and SD		
	AA	AS	SS
Total erythrocytes (10^6/ µl)	4.32±1.02	4.61±0.561	2.44±+0.94 *
Haemoglobin (g/dl)	12.31±2.80	12.50±1.98	6.59±2.43 *
Packed cell volume (PCV) or Haematocrit (Hct) %	39.94±8.12	39.86±5.39	22.03±11.91 *
Mean cell haemoglobin concentration (MCHC) g/dl	30.73±1.63	31.33±1.41	29.81±1.74
Mean cell volume (MCV) fl	89.04±7.71	86.55±5.99	92.51±11.91
Mean cell haemoglobin (MCH) pg	27.43±3.07	27.15±2.69	26.49±4.03
The total leukocytes (10^3/µl)	6.59±7.23	7.12±3.71	20.41±12.89 *
Platelet count (10^3/µl)	220.97±102.15	248.38±109.89	280.79±121.16 *

reported that, blood pictures in sickle cell trait group and normal individuals were normocytic normochromic and microcytic hypochromic .The total leukocytes count was significantly elevated in sickle cell disease when compared with normal individuals also total and segmented leukocyte numbers were greatly increased during vasoocclusive crisis and ininfection, but only with bacterial infection was there a consistent increase in bands or non-segmented leukocytes (mean, 4,580/µl). On the basis of these data we believe that total and differential leukocyte counts are of value for identifying those children with potentially serious bacterial infections,Buchanan GR&Glader BE ,(1978).

Despite a clear role for leukocytes in modulating the pathophysiology of sickle cell disease (SCD), the mechanism by which leukocyte numbers are increased in this disorder remains unclear. Hypothesizing that the chronic inflammatory state, elicited by adhesive interactions involving various cell types, might underlie leukocytosis,Conran N,et.al.,(2007). The platelet count was significantly increased in sickle cell disease patients suggesting the contribution of platelets in the vasoocclusive phenomena found in sickle cell anaemia, Ibanga IA,(2006).Sickle cell diseases is a genetic abnormality involving the haemoglobin , although, it is primarily a red cell disorders, the white blood cells and platetlets are also affected by the mutation,Akinsegun A.,et.al.,(2012). The characteristics of Hb SS disease observed in this study did not differ significantly from those of other studies in Nigeria,Nduka, N,et.al.,(1993), frequencies lower than that were reported in Bahrain, Sultanate of Oman and Turky in which SCT frequencies were 7%. 10% and 0.5% respectively,Buhazza MA, et.al.,(1985) ; Al Arrayed SS,et . al.,(1995).

The distribution of the S gene among various ethnic and linguistic groups in the Sudan studied,Mohammed AO A ,(2006),they found that, the majority of patients (93.7%) belonged to families of single ethnic descent, indicating the high degree of within-group marriages and thus the higher risk of augmenting the gene.

Some factors were acting to reflect high frequency of SCT in Sudan. These factors include illiteracy, consanguineous marriage, closure societies and lack of medical counseling. In order to avoid risk of having SCD, serious efforts are needed to improve social culture and economical status to help reducing this vast spreading and getting rid of sickle cell disease by reducing the gene distribution in future generation.

CONCLUSION

The frequencies of sickle cell trait and sickle cell diseases was higher among the participants and patients of sickle disease showed lower values of red blood cells parameters , but higher values of white blood cells and platelets compared to haemoglobin phenotype AA control participants.

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