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Frequency of Sickle Cell Trait among Relatives of Sickle Cell Anemia Patients in Al-Gadaref State-Sudan

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Abstract

Background: The term "sickle cell disease" refers to a collection of autosomal recessive genetic disorders characterized by the Hb S variant of the B-globin gene. Sickle cell disease is a major public health concern that has a great impact on both individuals and societies. In Sudan, sickle cell anemia is one of the major types of anemia, especially western Sudan, where the sickle cell gene is frequent, so this study aimed to determine the frequency of sickle cell trait (HbAS) among relatives of sickle cell anemia patients (HbSS) in Al-Gadaref state – Sudan.

Methodology: A descriptive, cross-sectional, analytical study was carried out for seventeen families with one hundred and fourteen individuals with different ethnic descents. 56 Males and 58 Females, age ranged between 1 -70 years compared with 30 healthy individuals age ranged between 28-73 as a control group. Venous blood (2.5ml) was collected from each individual in an ethylene diamine tetra acetic acid (EDTA) container for complete blood count (CBC), erythrocyte sedimentation rate (ESR), sickling test, and hemoglobin electrophoresis.

Results: The data showed that (67%) of the study population were positive and (33%) were negative for sickling test, the hemoglobin electrophoresis showed high frequency of (HbAS) (66.7%), normal people (HbAA) (24.6%), HbSS (5.3%), hemoglobin C trait (HbAC) (1.8%), and sickle cell with hemoglobin C disease (HbSC) (1.8%). The mean of Hb level, TRBCs, and PCV in patients with HbSS and HbSC were lower than in HbAS and HbAC. The MCV, MCHC, and MCH, showed no significant difference between different groups. The total leukocytes, was significantly elevated in HbSS and HbSC. Platelets were higher in HbSS and lower in HbSC, and ESR was elevated in both as compared with other groups.

Conclusion: The sickle cell trait is highly frequent among the relatives of sickle cell anemia patients and the spreading degree could be due to the high degree of consanguineous marriage in the studied population.

Introduction

Sickle cell trait (SCT) is a benign condition with no anemia and normal appearance of red blood cells on blood film. It is remarkably common in some parts of the world, carriers have a mixture of sickle cell hemoglobin and normal hemoglobin, and their erythrocytes do not sickle in vivo, so they have no hematological abnormality (A. C. Allison, 1954). Nevertheless, under unusual circumstances, serious morbidities or mortalities can result from complications related to polymerization of deoxy-hemoglobin S. (Niton Jon, 2010). Sickle cell trait affects 8 – 10% of African - Americans and up to 25 – 30% of the population in West Africa. It reduces the risk of severe falciparum malaria, but not the prevalence of parasitaemia. There appears to be no any effect on infections with other forms of malaria. (A victor Hoffbrand and et al, 2011).

Sickle cell traits present with varied problems, including increased urinary tract infections in women, gross hematuria, complications of hyphema, splenic infarction with altitude hypoxia or exercise, life threatening complications of exercise or idiopathic sudden death. (Sears DA, 1978), (Eichner ER, 2007). People with the uncomplicated sickle cell trait have a normal blood examination as assessed by conventional clinical methods, including normal red cell morphology, indices, reticulocytes count, and red blood cell survival by chromium labeling. Conventional methods of detecting hemolysis are negative, such as measurements of serum

Haptoglobin, bilirubin, and lactate dehydrogenase (LDH). Erythrocyte density distribution is normal, adherence to endothelium is not increased, altered membrane lipids and proteins are not detectable. Cytoplasmic inside-out vesicles with high calcium contents are absent, and permanently distorted erythrocytes are not observed. (Niton Jon, 2010).

HbC is found among individuals of African descent and the compound heterozygote state HbSC accounts for 25 – 50% of patients with SCD.The vaso - occlusive complications seen in patients with HbSC resemble those seen in patients with HbSS but are less severe (A Victor Hoffbrand and et al, 2011). Hemoglobin C trait describes the heterozygous condition in which there is one normal β^A gene and one abnormal β^C gene. It is of no clinical significance, but is of importance in counseling prospective parents. This is largely because of the possibility of sickle cell/hemoglobin C disease if one parent has the hemoglobin C trait and the other has sickle cell trait. (Barbara J. Bain, 2006).

In Sudan sickle cell anemia is one of the major types of aneamia, especially western Sudan, where the sickle cell gene is frequent (Abdelrahim O. et. al, 2006). The sickle cell gene may have been preferentially introduced through males of migrating West African tribes to Sudan, particularly Hosa, Folani, and Bargo (Rehab E. Bearer. et.al, 2007). The first report of the presence of the HbS gene in the Sudanese appeared in 1950 (Abbott PH, 1950). Later it was shown that the frequency of the gene varies significantly in different tribes (Foy H, Kondi A, 1954). The frequency of sickle cell trait has not been studied satisfactorily in Sudan, especially western and eastern parts where the gene frequency of sickle cell disease is quite prevalent. The problem becomes augmented due to population unawareness, consanguineous marriage, which employ widely in that area, lack of health counselling and undertaken serious researches (Munsoor M .et al, 2011). Therefore, this study was conducted in Algedaif state-Sudan in order to decrease the spreading of sickle cell disease in that area.

Material and Methods

A descriptive, cross-sectional, analytical study was carried out during the period of April –August 2012, to detect the SCT among relatives of sickle cell anemia patients in Al-Gadaref state - Sudan. The sampling method was a non probability sample calculated to achieve 114 specimens of blood. The sample size was determined according to available resources and facilities.

Sicklers' relatives were selected randomly regardless of their age or sex, prior to the study sociodemographic data which included (age, sex, father's tribe, mother tribe, area of origin and history of sickness) were obtained with consent signed by each individual .The inclusion criteria were all families having at least a confirmed patient diagnosed as having sickle cell anaemia (HbSS).

Venous blood of 2.5 ml was collected in EDTA container from all enrolled Sicklers' relatives and 30 healthy individuals as a control group. Data were analyzed using (SPSS) computer program.

Results:

The data showed that the gender enrolled in the study as in table (1). From the results of Hb electrophoresis its clear, that the highest frequency among the study population occur in HbAS (66.7%), followed by Hb AA (24.6%), HbSS (5.3%), and the lowest frequency among the study population is (1.8%) in HbAC, and HbSC, table (2). Table (3) also showed the Hb electrophoresis among the different tribes enrolled in the study.

Table (1): The gender, frequency among the study population.

Gender	Frequency	Present
Male	56	49.1
Female	58	50.9
Total	114	100.0

Table (2): The hemoglobin electrophoresis among study population

	Frequency	Present
A/A	28	24.6
A/C	2	1.8
A/S	76	66.7
S/C	2	1.8
S/S	6	5.3
	114	100.0

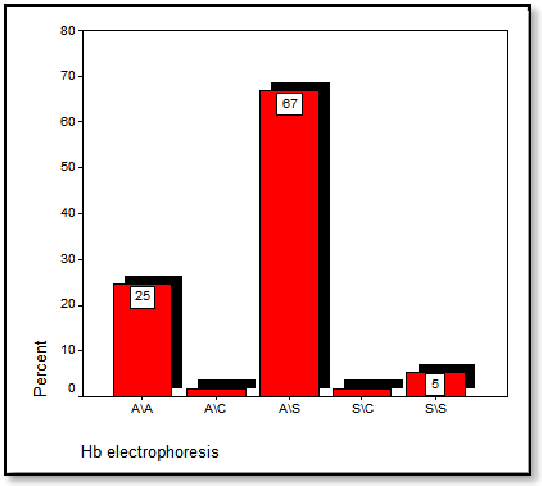
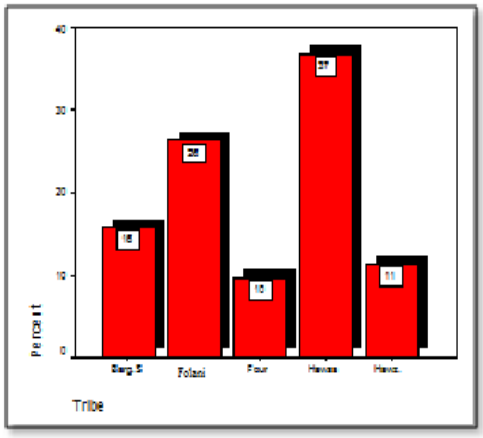


Table (3): The frequency distribution among the study population.

Tribe	Frequency	Present
Barg.S	18	15.8
Folani	30	26.3
Four	11	9.6
Hawsa	42	36.8
Hawz	13	11.4
	114	100.0



The statistical analysis of the data for mean, standard deviation as shown in table (4-7) for complete blood count (CBC), and erythrocyte sedimentation rate (ESR) in different groups of HbAS, HbAC, HbSC and HbSS, as compared to control group (HbAA).

Table (4): Complete blood count and ESR in HbAA, and HbAC groups. (The mean of ESR in mm/h, platelets in $x10^3$/µl, MCHC in mg/dl, MCH in pg, MCV in FL, PCV in %, Hb in g/dl, RBC in $x10^6$/ µl, WBC in $x10^3$/µl).

Group Statistics

	Hb electrophoresis	N	Mean	Std. Deviation	P. value
ESR	A\A	28	11.5000	6.58562	.483
	A\C	2	8.0000	9.89949	
platelet	A\A	28	237.50	42.568	.938
	A\C	2	235.00	63.640	
MCHC	A\A	28	32.3571	2.34006	.665
	A\C	2	33.1000	1.55563	
MCH	A\A	28	28.1679	2.17205	.598
	A\C	2	29.0000	.00000	
MCV	A\A	28	83.6429	7.18132	.342
	A\C	2	88.6500	3.32340	
PCV	A\A	28	37.36	4.399	.267
	A\C	2	41.00	4.243	
Hb	A\A	28	12.439	1.0884	.189
	A\C	2	13.500	.7071	
RBC	A\A	28	4.432	.4691	.625
	A\C	2	4.600	.2828	
WBC	A\A	28	4.536	1.7498	.465
	A\C	2	3.600	.8485	
Age	A\A	28	15.14	12.753	.095
	A\C	2	31.00	1.414	

Table (5): Complete blood count and ESR in HbAA, and HbAS groups.(The mean of ESR in mm/h, platelets in $x10^3$/µl, MCHC in mg/dl, MCH in pg, MCV in fl, PCV in %, Hb in g/dl, RBC in $x10^6$/ µl, WBC in $x10^3$/µl).

Group Statistics

	Hb electrophoresi	N	Mean	Std. Deviation	P.value
ESR	A\A	28	11.5000	6.58562	.000
	A\S	76	21.6974	13.53122	
platelet	A\A	28	237.50	42.568	.530
	A\S	76	230.71	50.828	
MCHC	A\A	28	32.3571	2.34006	.292
	A\S	76	32.7737	1.52686	
MCH	A\A	28	28.1679	2.17205	.229
	A\S	76	27.5197	2.50902	
MCV	A\A	28	83.6429	7.18132	.680
	A\S	76	82.9250	8.08594	
PCV	A\A	28	37.36	4.399	.513
	A\S	76	38.20	6.223	
Hb	A\A	28	12.439	1.0884	.860
	A\S	76	12.508	1.9402	
RBC	A\A	28	4.432	.4691	.173
	A\S	76	4.601	.5857	
WBC	A\A	28	4.536	1.7498	.193
	A\S	76	5.136	2.1772	
Age	A\A	28	15.14	12.753	.007
	A\S	76	25.20	17.679	

Table (6): Complete blood count and ESR in HbAA, and HbSC groups.(The mean of ESR in mm/h, platelets in $x10^3$/µl, MCHC in mg/dl, MCH in pg, MCV in fl, PCV in %, Hb in g/dl, RBC in $x10^6$/ µl, WBC in $x10^3$/µl).

Group Statistics

	Hb electrophoresis	N	Mean	Std. Deviation	P.value
ESR	A\A	28	11.5000	6.58562	.000
	S\C	2	95.0000	7.07107	
platelet	A\A	28	237.50	42.568	.017
	S\C	2	160.00	14.142	
MCHC	A\A	28	32.3571	2.34006	.705
	S\C	2	33.0000	.00000	
MCH	A\A	28	28.1679	2.17205	.295
	S\C	2	26.5000	.70711	
MCV	A\A	28	83.6429	7.18132	.376
	S\C	2	79.0000	1.41421	
PCV	A\A	28	37.36	4.399	.005
	S\C	2	27.50	3.536	
Hb	A\A	28	12.439	1.0884	.000
	S\C	2	9.050	1.3435	
RBC	A\A	28	4.432	.4691	.006
	S\C	2	3.400	.5657	
WBC	A\A	28	4.536	1.7498	.004
	S\C	2	8.500	.7071	
Age	A\A	28	15.14	12.753	.328
	S\C	2	6.00	4.243	

Table (7): Complete blood count and ESR in HbAA, and HbSS groups. (The mean of ESR in mm/h, platelets in $x10^3$/µl, MCHC in mg/dl, MCH in pg, MCV in fl, PCV in %, Hb in g/dl, RBC in $x10^6$/ µl, WBC in $x10^3$/µ).

Group Statistics

	Hb electrophoresi	N	Mean	Std. Deviation	P.value
ESR	A\A	28	11.5000	6.58562	.000
	S\S	6	42.3333	11.84342	
platelet	A\A	28	237.50	42.568	.000
	S\S	6	376.67	13.663	
MCHC	A\A	28	32.3571	2.34006	.308
	S\S	6	33.3667	.66533	
MCH	A\A	28	28.1679	2.17205	.924
	S\S	6	28.2667	2.83314	
MCV	A\A	28	83.6429	7.18132	.395
	S\S	6	86.5000	8.31264	
PCV	A\A	28	37.36	4.399	.000
	S\S	6	22.67	2.875	
Hb	A\A	28	12.439	1.0884	.000
	S\S	6	7.483	.9948	
RBC	A\A	28	4.432	.4691	.000
	S\S	6	2.700	.2280	
WBC	A\A	28	4.536	1.7498	.000
	S\S	6	12.167	.7528	
Age	A\A	28	15.14	12.753	.448
	S\S	6	11.00	6.356	

Discussion:

Sickle cell disease is a major public health concern that has a great impact on both individuals and societies. In Sudan, sickle cell anemia is one of the major types of anemia, especially western Sudan, where the sickle cell gene is frequent.The frequency of sickle cell trait in our study was higher as compared with other results in Nigeria and Saudia Arabia that reported by Ambe JP. et.al 2012, in Borno and Yobe State that had the highest percentage of sickle cell trait in Nigeria with prevalence of 27.9% and 32.6% respectively, and the result of Wasil Jastaniah 2011, reported that the prevalence of sickle-cell trait ranges from 2% to 27%, in some areas of Saudi Arabia. Our study was within range of the study done in western area of Sudan that showed the frequency of sickle cell trait in the study area was 54 which means that 54% of the studied population were carriers of sickle cell gene (Munsoor M and Afaf A, 2011).

The high frequency of sickle cell disease is higher among Hawsa (36.8%), followed by Folani (26.3%), Bargo Selehab (15.8%), Hawzma (11.8%), and lastly Four (9.6%). (79%) of study population their tribes were originally from Afro-Asiatic origin and (21%) their origin from western area of the Sudan. No any case of sickle cell anemia was detected from eastern, northern or southern tribes of the Sudan. The study of Abderahim O. and Attalah B, showed sickle cell anemia was found to be predominant among the Afro-Asiatic-speaking groups (68.4%) including nomadic groups of Arab and non- Arab descent that migrated to the Sudan in various historical epochs. Its also similar to study done by Omer, etal 1972 about the abnormal haemoglobins in the indigenous and immigrant tribes of the Sudan that showed the highest sickle cell trait incidence was found in the immigrant tribes.The majority of the study population (98.24%) belonged to families of single ethnic descent, which reflected the high degree of within -group marriage thus in a high risk of augmenting the sickle cell gene.

Erythrocyte sedimentation rate in the study population is significantly higher in HbSS and HbSC people. This may be due to any type of infections or other causes because the erythrocyte sedimentation rate of asymptomatic patients with sickle cell anemia is abnormally low and in patients with sickle crisis and medical complications the sedimentation rates were even higher (Lawrence C, Fabry ME,1986). Also the higher ESR was found in HbAS than in HbAA this may be related to their older age or having any other cause that increased their ESR.

The mean of Hb level, TRBCs, and PCV in patients with sickle cell anemia and HbSC are lower than in sickle cell trait and HbAC which are not significantly differ than normal person HbAA, These results were match with (Akinsegun A, Adedoyin D, 2012) and (Hoff brand and Bettit, 1993), who showed the reduction of the above values in HbSC, HbSS, and no significant change between HbAA, HbAS, and HbAC. The result of MCV, MCHC, and MCH, showed no significant difference between all electrophoresis groups, these results were in agreeing with the study of A. Vector Hoffbrand. et.al, 2005, that showed sickle cell anemia was due to normocytic normochromic anemia, and carriers were asymptomatic, which had normal red cell indices. Similar

studies of Serjeant GR and Serjeant BE, 1972, also showed that compound heterozygote (HbSC), individuals their MCV and MCH were lower as compared to sickle cell anemia individuals.

The leukocytes were elevated in HbSS and HbSC and normal in HbAC, HbAS, and HbAA, similar results were obtained from study of Akinsegun A, Adedoyin D, 2012, concluded that the higher values of white cell count and platelets in sickle cell anemia as compared to hemoglobin phenotype AA controls. Also the study of Wong W-Y, Zhou Y, et al. 1996, showed that the WBC, neutrophil count and monocyte count were elevated in sickle cell hemoglobin C disease, but less than in sickle cell anemia. Similar results were published in the study of Malik H.I.M, et.al.2013, for the frequency of sickle cell disease in the Heglig area in Sudan .

The platelet count was higher in HbSS that agree with Akinsegun A and Adedoyin D and lower in HbSC as a result of Splenic sequestration that associated not only with a fall in the haemoglobin concentration, but also with a fall in the platelet count (Zimmerman SA, Ware RE, 2000), no significant change between platelet values in HbAA, HbAS and HbAC.

From the present study it concluded that sickle cell trait was highly frequent among the relatives of sickle cell anemia patients in the study area and could be capable of spreading the disease further due to the high degree of consanguineous marriage, population unawareness, closure societies and lack of medical counseling, and provide that sickle cell anemia was found to be predominant among afro- Asiatic speaking groups.

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