Association between Sick Cell Crisis and Other Types of Hemoglobin among Sudanese Children with Sick Cell Anemia

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Abstract

Background: Sick cell anemia (SCA) is a common genetic disorder that causes considerable morbidity and mortality throughout the world. The study was performed in 82 Sudanese participants with sickle cell disease as study population with both sexes, age (2-12 years). **Materials and Methods:** Venous blood was collected using sterile disposable plastic syringe after cleaning the vein puncture area with 70% ethanol, the blood was added to the EDTA and gently mixed, then CBC was performed using (Sysmex-3000 plus) and (Electrophoresis) was done. **Results:** The results of this study show that most patients had raised level of hemoglobin F according to gender found that there were 43 males and 28 females had raised level of HbF, 7 males and 2 females were normal, and 36% of patients had raised level of HbA2 in both gender (20 males) (10 females). **Conclusion:** HbF in sickle cell anemia patients was found to be 85.3% (raised), HbS in sickle cell anemia patients was found to be 100% (raised) and HbA2 in sickle cell anemia patients was found to be 30.5% (Normal).

Keywords

Sudanese Children, Sick Cell Crisis, other Types of Hemoglobin

1. Introduction

Sickle cell anaemia (SCA) is a common genetic disorder that causes considerable morbidity and mortality throughout the world. SCA an amino acid substitution of valine for glutamic acid at position 6 of the β -globin chain, which results in the polymerization of haemoglobin upon DE oxygenation, leading to deformed dense red blood cells. The predominant pathophysiological feature of SCA is vaso-occlusion, which leads to acute and chronic complications such as painful crises, acute chest syndrome and strokes. Patients with SCA have a markedly decreased life expectancy and their quality of life is greatly compromised by their disease [1] [2]. In 1910, Dr James Herrick working in Chicago, USA, reported "Peculiar elongated and sickle shaped red blood corpuscles in a case of severe anaemia" [2] [3] [4]. The inherited disease was subsequently called SCA, and has continued to attract the attention of medical scientists to the present day. SCA includes homozygous (Hb SS) sickle cell disease and compound heterozygous states such as sickle cell haemoglobin C (Hb SC) disease, sickle cell thalassemia [5] [6] [7]. Haemoglobin S becomes polymerized and becomes poorly soluble when the oxygen tension is lowered and red

cells that contain this haemoglobin become distorted and rigid. SCD occurs when an individual is homozygous for the sickle cell mutation or is a compound heterozygote for sickle haemoglobin and β -thalassemia [8] [9] [10].

2. Materials and Methods

Study Participants: The study was performed in 82 Sudanese patients with sickle cell disease as study population with both sexes, age (3-12 years).

Sample preparation and methods:

Venous blood was collected using sterile disposable plastic syringe after cleaning the vein puncture area with 70% ethanol, the blood was added to the EDTA and gently mixed.

Automated hematology analyzer (BC-3000 plus) technique:

The Coulter principle is based on the following: particles suspended in an isotonic diluent, when drawn through an aperture which has an electric current flowing through it was caused a measurable drop in voltage which is proportional to the size of the particle passing through the aperture is constant the particle can be quantified per unit volume. This is also called electrical impedance.

Hemoglobin electrophoresis:

The CAPILLARYS 2 FLEX-PIERCING instrument uses the principle of capillary electrophoresis in free solution. With this technique, charged molecules are separated by their electrophoretic mobility in an alkaline buffer with a specific pH. Separation also occurs according to the electrolyte pH and electro osmotic flow.

Statistical Analysis:

Statistical assessment was carried out with statistical package for social sciences (SPSS) version 17.0 for windows statistical software.

3. Results

The results were compared according to the objectives of the study as illustrated below: Table 1 shows the distribution of gender among study population (51) of patients (62.2%) are male, (31) of patients are female (37.8%), Table 2 shows distribution of age among the study population into two groups (2-7) 50%, (8-12) 50%, Table 3 shows distribution of hemoglobin according to gender and severity of anemia, found 44 male suffering from severe anemia and 7 male moderate anemic, 82 females suffering from severe anemia and 3 females were moderate anemic, Table 1 shows distribution of red blood cell according to gender found that 49 males and 31 females had erythropenia, 2 males were normal, Table 2 shows distribution of white blood cell according to gender found that 27 males and 23 females suffering from leukocytosis and 24 males and 8 females were normal, Table 3 shows distribution of platelets according to gender 27 males and 15 females had thrombocytosis, 3 males and 2 females had thrombocytopenia, and 21 males and 14 females were normal, Table 4 shows distribution of hemoglobin F according to gender found 43 males and 28 females had raised level of HbF, 1 male and 1 female had decreased level of HbF, 7 males and 2 females were normal. Table 5 shows distribution of hemoglobin A2 according to gender found that 20 males and 10 females had raised level of HbA2, 1 female had decreased level of HbA2, 31 males and 20 females were normal, Table 6 shows distribution of hemoglobin according to age found that (2-7 years) 35 had severe anemia and 6 had moderate anemia, and in the other age group (8-12 years) 37 had severe anemia and 4 had moderate anemia, Table 7 shows distribution of red blood cell according to gender found that in both age group (2-7 years) and (8-12 years) 40 had erythropenia and 1 was normal. Table 8 shows distribution of white blood cell according to age found that in both age groups (2-7 years) and (8-12) 25 patients had leukocytosis and 16 were normal. Table 9 shows distribution of platelets according to age found that age group (2-7years) 22 patients had thrombocytosis, 2 patients had thrombocytopenia and 17 patients were normal, the other age group (8-12 years) 20 patients had thrombocytosis, 3 patients thrombocytopenia and 18 were normal. Table 10 shows distribution of HbF according to age found that the age group (2-7 years) 33 patients had raised HbF level, 2 patients had decreased level of HbF and 6 patients were normal, in the other age group (8-12years) 38 patients had raised level of HbF and 3 patients were normal. Table 11 shows distribution of HbA2 according to the age found that the age group (2-7 years) there was 15 patients had raised HbA2 level and 26 patients were normal in the other age group (8-12 years), there was 15 patients had raised HbA2 level, 1 patient had decreased HbA2 level and 25 patients were normal. Table 12 shows correlation between hemoglobin and red blood cells found that in 72 patients that had severe anemia there was 71 patients had erythrocytopenia and 1 patient was normal, and in 10 patients that had moderate anemia there was 9 patients had erythrocytopenia and 1 patient was normal. Table 13 shows comparison between hemoglobin and white blood cells found that in 72 patients that had severe anemia there was 47 patients had leukocytosis and 25

patients were normal, and in 3 patients that had moderate anemia there was 9 patients had leukocytosis and 7 patients were normal.

Table 14 shows correlation between hemoglobin and platelets found that in 72 patients had severe anemia there was 36 patients had thrombocytosis, 4 patients had thrombocytopenia and 32 were normal, and in 10 patients that had moderate anemia there was 6 patients had thrombocytosis, 1 patient had thrombocytopenia and 3 patients were normal. Table 15 shows comparison between hemoglobin and HbF found that in 72 patients that had severe anemia there was 62 patients had raised HbF level, 2 patients had decreased HbF level and 8 patients were normal, and in 10 patients that had moderate anemia there was 9 patients had raised HbF level and 1 patient was normal. Table 16 shows comparison between hemoglobin and HbA2 found that in 72 patients that had severe anemia there was 28 patients had raised HbA2 level, 1 patient had decreased HbA2 level and 43 patients were normal, and in 10 patients that had moderate anemia there was 2 patients had raised HbA2 level and 8 patients were normal. Table 17 shows mean and SD of Hb F, Hb S and HbA2 male (2.71 ± 0.701) female (2.84 ± 0.523), (2.0 ± 0.0) for both genders and male (1.78 ± 0.986) female (1.68 ± 0.945) respectively.

Figure 1 shows scatter plot of correlation between hemoglobin% and HbF%, Figure 2 shows scatter plot of correlation between hemoglobin% and HbS%, Figure 1 shows scatter plot of correlation between hemoglobin% and age, Figure 2 shows scatter plot of correlation between hemoglobin% and HbA2%.

Table 1. Distribution of Gender among study population

	Frequency	Percent
male	51	62.2
female	31	37.8
Total	82	100.0

Table 2. Distribution of the age among the study population

	Age	
	Frequency	Percent
2-7	41	50.0
8-12	41	50.0
Total	82	100.0

Table 3. Distribution of hemoglobin according to gender

gender * Hemoglobin				
Count				
		Hemoglobin %		Total
		severe anemia	anemia	
Gender	Male	44	7	51
	female	28	3	31
Total		72	10	82

Table 4. Distribution of red blood cell according to gender

gender * red blood cells				
		Count		
		red blood cells		Total
		4-5.5	less than 4	
Gender	Male	2	49	51
	female	0	31	31
Total		2	80	82

Table 5. Distribution of white blood cell according to gender

gender * white blood cells				
		Count		
		white blood cells		Total
		4.5-14.5	more than 14.5	
Gender	male	24	27	51
	female	8	23	31
Total		32	50	82

Table 6. Distribution of platelets according to gender

gender * platelets					
		Count			
		Platelets			
		150-400	less than 150	more than 400	Total
Gender	Male	21	3	27	51
	Female	14	2	15	31
Total		35	5	42	82

Table 7. Distribution of hemoglobin F according to gender

gender * Hemoglobin F%					
		Count			
		Hemoglobin F%			Total
		1%-2%	less than1%	more than2%	
Gender	Male	7	1	43	51
	female	2	1	28	31
Total		9	2	71	82

Table 8. Distribution of hemoglobin A2 according to gender

gender * Hemoglobin A2%					
		Count			
		Hemoglobin A2%			Total
		2%-3%	less than 2%	more than 3%	
Gender	Male	31	0	20	51
	female	20	1	10	31
Total		51	1	30	82

Table 9. Distribution of hemoglobin according to age

age * Hemoglobin%				
		Count		
		Hemoglobin%		
		severe anemia	anemia	Total
Age	2-7	35	6	41
	8-12	37	4	41
Total		72	10	82

Table 10. Distribution of red blood cell according to age

age * Red blood cells				
		Count		
		Red blood cells		Total
		4-5.5	less than 4	
Age	2-7	1	40	41
	8-12	1	40	41
Total		2	80	82

Table 11. Distribution of white blood cell according to age

age * white blood cells				
		Count		
		white blood cells		Total
		4.5-14.5	more than 14.5	
Age	2-7	16	25	41
	8-12	16	25	41
Total		32	50	82

Table 12. Distribution of platelets according to age

age * platelets					
		Count			
		Platelets			
		150- 400	less than 150	more than 400	Total
Age	2-7	17	2	22	41
	8-12	18	3	20	41
Total		35	5	42	82

Table 13. Distribution of hemoglobin F according to age

age * Hemoglobin F%					
		Count			
		Hemoglobin F%			
		1%-2%	less than1%	more than2%	
Age	2-7	6	2	33	41
	8-12	3	0	38	41
Total		9	2	71	82

Table 14. Distribution of hemoglobin A2 according to age

age * Hemoglobin A2%					
		Count			
		Hemoglobin A2%			
		2%-3%	less than 2%	more than 3%	Total
Age	2-7	26	0	15	41
	8-12	25	1	15	41
Total		51	1	30	82

Table 15. Comparison between hemoglobin and red blood cell

Hemoglobin % * Red blood cells				
		Count		
		Red blood cells		Total
		4-5.5	less than 4	
Hemoglobin%	severe anemia	1	71	72
	anemia	1	9	10
Total		2	80	82

Table 16. Comparison hemoglobin and white blood cell

Hemoglobin % * white blood cells				
		Count		
		white blood cells		Total
		4.5-14.5	more than 14.5	
Hemoglobin%	severe anemia	25	47	72
	Anemia	7	3	10
Total		32	50	82

Table 17. Comparison between hemoglobin and platelets

Hemoglobin % * platelets					
		Count			
		Platelets			Total
		150-400	less than 150	more than 400	
Hemoglobin%	severe anemia	32	4	36	72
	anemia	3	1	6	10
Total		35	5	42	82

Table 18. Comparison between of hemoglobin and hemoglobin F

Hemoglobin % * Hemoglobin F%					
		Count			Total
		Hemoglobin F%			
		1%-2%	less than1%	more than2%	
Hemoglobin%	severe anemia	8	2	62	72
	anemia	1	0	9	10
Total		9	2	71	82

Table 19. Comparison between hemoglobin and hemoglobin A2

Hemoglobin % * Hemoglobin A2%					
		Count			
		Hemoglobin A2%			Total
		2%-3%	less than 2%	more than 3%	
Hemoglobin%	severe anemia	43	1	28	72
	anemia	8	0	2	10
Total		51	1	30	82

Table 20. Mean and standard deviation of different hemoglobin types

Group Statistics					
	gender	N	Mean	Std. Deviation	Std. Error Mean
Hemoglobin F%	male	51	2.71	0.701	0.098
	female	31	2.84	0.523	0.094
Hemoglobin S%	male	51	2.00	0.000 ^a	0.000
	female	31	2.00	0.000 ^a	0.000
Hemoglobin A2%	male	51	1.78	0.986	0.138
	female	31	1.68	0.945	0.170

a. It cannot be computed because the standard deviations of both groups are 0.

Figure 1. Scatter plot of correlation between hemoglobin% and HbF%.

Figure 2. Scatter plot of correlation between hemoglobin% and HbS%.

Figure 3. Scatter plot of correlation between hemoglobin% and age.

Figure 4. Correlation between hemoglobin and HbA2.

4. Discussion

All around world continuous research is conducting in physio chemicals of anticoagulant prosperities, these study well thought out to therapeutically better anticoagulant, multiple targets and safer. This study the mean and standard deviation of HbF, HbS and HbA2 in percentages (2.76 ± 0.639), (2.0 ± 0.0) and (1.74 ± 0.966) respectively in our study. The mean of Hb S is the larger indicate that most patients have large level of Hb S which is the diagnostic hemoglobin for sickle cell anemia. The mean of HbF is increased and HbA2 is also increased. Hb F, Hb S and

HbA2 show different means and standard deviations in males and females as HbF% in males (2.71 ± 0.701) while in females (2.84 ± 0.523) P value (0.098) for males and (0.094) for females, HbA2 (%) in males (1.78 ± 0.0986) while in females (1.68 ± 0.945) P value (0.138) for males (0.170) for females, but similar in HbS% in males (2.0 ± 0.0) and in females (2.0 ± 0.0) P value (0.0) for both sex. Our study agree with study done by [11] in Pakistan it was concluded that The mean of Hb S is the larger indicate that most patients have large level of Hb S which is the diagnostic hemoglobin for sickle cell anemia and there is the mean of HbF is increased and HbA2 is also increased. Hb F, Hb S and HbA2 show different means and standard deviations in males and females as HbF% in males (2.71 ± 0.701), also agreed with study done by [12] at 2002 in Sudan was found sickle cell anemic patient HbF level about (7+ or -5.6%), while sickle cell trait patient had (0.8+ or -0.5) and sickle cell HbS F was found Hb F level (17.4+ or -4.2%) in other research sickle cell anemic patient had (9.2+ or -4.9%) of HbF level. As reported by this research that sickle cell anemic patient in their first decade of life (0-10 years) had higher HbF (8.2+ or -5.8) than patient in old age group (above 10 years) HbF level ($2.7 +$ or -1.1%).

5. Conclusion

This study concluded the HbF in sickle cell anemia patients was found to be 85.3 (raised), the HbS in sickle cell anemia patients was found to be 100% (raised) and the HbA2 in sickle cell anemia patients was found to be 30.5% (Normal).

List of abbreviations

SCA—Sickle Cell Anaemia
SCD—Sickle Cell Disease
HB-SS—Homozygous Haemoglobin of Sickle Cell
HB-SC—Heterozygous Sickle Cell of Haemoglobin C
DNA—Deoxy Ribo Nucleic Acid
VCAM—Vascular Cell Adhesion Molecule
IgG—Immunoglobulin G
NO—Nitric Oxide
HB-A—Adult Haemoglobin
2,3-DPG—2,3-diphosphoglycerate
MCV—Mean Corpuscular Volume
MCH—Mean Corpuscular haemoglobin
HB-F—Fetal haemoglobin
HBFH—Hereditary Persistence Fetal Haemoglobin
EDTA—Ethylene Diamine Tetra Acetic Acid
CBC—Complete Blood Count
SPSS—Statistical Package of Social Sciences
SD—Standard Deviation

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