Glucose 6 Phosphate Dehydrogenase Deficiency Screen among Males Patients with Sickle Cell Disorders in Sudan

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Abstract

The intent of this study is screening patients with both Sickle Cell anemia and Sickle Cell trait for Glucose 6 Phosphate Dehydrogenase deficiency and to the effect of the combined disease on Red Cell Indices and Leukocytes in such patients.

The study was performed in Ja'afer Ibn-Aof Pediatric Hospital (C_{15}) and National Health Laboratory (STAC) from April 20 to June 6, 2009. Blood samples were collected from 40 male patients with both types of Sickle Cell abnormalities (10 from AS and 30 from SS) and subjected for Methemoglobin Reduction test and Full Blood Count.

Results showed that G6PD deficiency was detected in 7 (23.3%) patients with Sickle cell anemia and precipitated insignificant decrease in all Red cell indices and an increase in White blood cell count.

The study concluded that G6PD deficiency result in the decrease of Red cell indices as well as the increase of White cell count in spite of statistical insignificance. Therefore, authors recommend screening all Sickle cell anemia patients for G6PD and special care should be vacant to them.

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Introduction

Sickle cell disorders are a group of genetic diseases of blood that expressed in individuals who are homozygous or heterozygous for the sickle hemoglobin (Hb-S) gene. Sickle Cell Anemia (SCA) is the most common inherited disease of hemoglobin consequent to inheritance of mutant hemoglobin genes from both parents [1,2] and it causes chronic hemolytic anemia, a recurrent, painful crisis, growth impairment and general ill health [3]. While Sickle Cell trait is inherited as mutant hemoglobin genes from one parent.

The pathogenesis occurs due to the polymerization of deoxygenated Hb-S as the formed polymer alters the normal biconcave disc shape into a rigid membrane, irregular sickled shape, unstable cell, causing intravascular hemolysis [4,5]. Moreover, hemolysis and recurrent Vaso-occlusion leads to chronic organ damage including the spleen, brain, bone and the penis [6].

Patients with sickle cell disease are highly susceptible to oxidative damage due to chronic redox imbalance in erythrocytes. Many authors indicated that sickle cell red cells generates twice as much as hydrogen peroxide, superoxide, and hydroxyl radicals [7,8,9], known as Reactive Oxygen Species, compared to normal healthy controls [10].

It therefore remains possible that Hb-SS patients with G6PD deficiency will be more prone to the development of accelerated hemolysis when exposed to Reactive Oxygen Species (ROS) that seem to aggravate hemolysis in Sickle Cell Anemia patients with G6PD deficiency [11].

The hemolysis of young erythrocytes that rich in G6PD, more likely complicate sickled cells complaint, particularly when these populations are shifted to the oldest cell during the aplastic crisis [12].

Therefore, objectives of this study are to determine the occurrence of G6PD deficiency among male patients with Sickle Cell abnormalities as well as to detect the effect of the combined disease on the severity of the illness in such patients.

Material and methods

A descriptive cross sectional study utilized qualitative approach was carried out during the period from 20/4/2009 to 4/6/2009 including 40 patients well diagnosed with sickle cell abnormality who are randomly selected.

Five-milliliter blood samples were withdrawn from each patient, collected in EDTA containers containing 0.04mg anticoagulant, mixed well and used within one hour. Thereafter, all blood samples were subjected for G6PD deficiency screen (Methemoglobin Reduction Test) as described by Dacie and Lewis [13] and Full Blood Count using Sysmex X-21 Auto-analyzer. Data was analyzed by the computer using SPSS statistical software.

Results

Table 1 Glucose 6 Phosphate screen in relation to Hemoglobin S variant

Hb S variant		**G6PD deficiency**		**Total**
		Negative	***Positive***	
Hb AS	***Number***	10	0	10
	%	25%	0%	25%
Hb SS	***Number***	23	7	30
	%	57.5%	17.5%	75%
Total	***Number***	33	7	40
	%	82.5%	17.5%	100%

P-value 0.093

As shown in table 1, Glucose 6 Phosphate deficiency detected in Sicklar patients with Hb SS genotype only, but without statistical significance.

Table 2 Hemoglobin level, PCV, Red Blood Cells count and White Blood Cells count in association to G6PD screen

G6PD Screen	Hemoglobin		PCV		Red B Cells		White B Cells	
	Mean	*SD*	*Mean*	*SD*	*Mean*	*SD*	*Mean*	*SD*
Positive	6.914	1.4029	22.471	4.4672	2.587	0.5506	25.729	11.9531
Negative	8.882	3.9352	27.418	10.4953	3.354	1.3845	21.248	13.6343
P. value	0.442		0.421		0.344		0.344	

Table 2 illustrates that Hemoglobin levels, PCV and red blood cell count were insignificantly decreased in Sicklar patients who are G6PD deficient. Unlike white blood cells count which is insignificantly increased in G6PD deficient Sicklar patients.

Table 3 Association between red cell indices and Glucose 6 Phosphate screen

G6PD Screen	MCV		MCH		MCHC	
	Mean	*SD*	*Mean*	*SD*	*Mean*	*SD*
Positive	86.857	5.0146	26.571	2.1769	30.600	1.7944
Negative	79.406	15.3234	26.161	5.24	31.639	3.3698
P. value	0.137		0.835		0.218	

Table 3 indicated the insignificant increase of MCV in Sicklar patients suffering G 6PD deficiency as well as the equal values of red cell indices in both deficient and non-deficient Sicklar patients.

Table 4 The association between G6PD screen and Jaundice, Bacterial Infection and previous transfusions

G6PD Screen		Jaundice		Bacterial Infection		Previous Transfusion	
		Present	*Absent*	*Present*	*Absent*	*Yes*	*No*
Negative	*Number*	10	23	9	24	13	20
	%	25%	57.5%	22.5%	60%	32.5%	50%
Positive	*Number*	3	4	3	4	5	2
	%	7.5%	10%	7.5%	10%	12.5%	5%
P-value		0.519		0.414		0.122	
Total	*Number*	13	27	12	28	18	22
	%	32.5%	67.5%	30%	70%	45%	55%

Table 4 indicates the insignificant statistical difference between G6PD screen and Jaundice, Bacterial infection nor pervious transfusions, although there is slight increase of the number of jaundice and bacterial infected sicklar patients as well as patients subjected for blood transfusions with negative G6PD than positive one.

Discussion

There is a high incidence of G6PD deficiency in patient with Sickle Cell Disease as it observed in a regions where Malaria is endemic particularly Falciparum Malaria which, lead many authors to suggest a protective role of G6PD on such patients [11,14].

In the present study, all Sickle Cell Trait patients showed normal G6PD enzyme efficiency. This finding is a proximal to 0.9% frequency of G6PD among Sickle cell trait patients reported by P. Heller *et al*, on hospitalized black male patients [15].

On the other hand, 23.3% of Sickle cell anemia patients showed G6PD deficiency, which is greater than that, obtained by Saad ST and Costa FF [16] who detected 9.7 % G6PD deficiency among Sickle cell disease patients in Brazil. While, Akbar M Mohammed and others [17] reported as high as 47% of Sickle cell anemia patients were recognized as G6PD deficient in Bahrain.

Although, the effect of G6PD enzyme deficiency on the decrease of Red Cell Indices and the increase of White blood cell count is observable however, it is of no statistical significance. Similar results detected by Baker H [18] in Saudi Arabia, Jane Wordle [19] in Jamaica and Saad ST and Costa FF [16] in Brazil.

The study concluded that G6PD deficiency was detected in 7 (23.3%) patients with Sickle cell anemia and result in the decrease of Red cell indices as well as the increase of White cell count in spite of statistical insignificance. Therefore, authors recommend screening all Sickle cell anemia patients for G6PD and special care should be vacant to them.

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