Assessment of Glucose-6-Phosphate Dehydrogenase Level among Sudanese Sickle Cell Anemia Patients

Ashraf Saleh Abd-Algadir Ahmed¹ and Mahdi H. A. Abdalla²*

¹Department of Hematology, Faculty of Medical Laboratory Sciences, Al neelin University, Khartoum, Sudan.
²Associate Professor of Hematology, Department of Hematology, Faculty of Medical Laboratory Sciences, Omdurman Ahlia University, Sudan.

*Corresponding Author: Mahdi H. A. Abdalla
Associate Professor of Hematology, Department of Hematology, Faculty of Medical Laboratory Sciences, Omdurman Ahlia University, Sudan.

ABSTRACT
Sickle cell disease is the most common haemoglobinopathy in Sudan, especially in western Sudan in which the sickle cell gene is frequent. Sickle cell disease leads to serious crises and complication, which considered life-threatening and fatal. The co-incidence of glucose-6-phosphate dehydrogenase deficiency and sickle cell disease may increase the severity of the disease. Patients with glucose-6-phosphate dehydrogenase deficiency may exhibit nonimmune hemolytic anemia in response to a number of causes, most commonly infection or exposure to certain medications or chemicals. Objectives: This study aimed to study the association of glucose-6-phosphate dehydrogenase (G6PD) deficiency and sickle cell disease in Sudan. Materials and Methods: This is a case-control study conducted in Khartoum, Sudan. It included 50 Sudanese Patients with sickle cell anemia, their G6PD levels was measured and compared with 50 age and sex matched normal subjects as control. G6PD level was determined by full automation (Mindary Bs 480), Haematological values were measured by Automated cell counter (symrex KX-21N). Results: This study investigated 50 cases and 50 controls. Their age was between 1-16 years, mean level of G6PD in cases (1808.6) was lower than controls (1831.8). Low G6PD level among cases showed strong positive correlation with RBCs, Hb and PCV. Conclusion: low G6PD level was found in sickle cell patients, the relationship of G6PD level was not significant when compared with the controls and positively correlated with RBCs, Hb, and PCV.

KEYWORDS: G6PD, sickle cell anemia, Sudan.

INTRODUCTION
The highest frequency of sickle cell disease is found in tropical regions, particularly sub-Saharan Africa, India and the Middle-East.¹ Migration of substantial populations from these high prevalence areas to low prevalence countries in Europe has dramatically increased in recent decades and in some European countries sickle cell disease has now overtaken more familiar genetic conditions such as hemophilia and cystic fibrosis.²

Three quarters of sickle-cell cases occur in Africa. A recent WHO report estimated that around 2% of newborns in Nigeria were affected by sickle cell anemia, giving a total of 150,000 affected children born every year in Nigeria alone. The carrier frequency ranges between 10% and 40% across equatorial Africa, decreasing to 1–2% on the North African coast and <1% in South Africa.³

In United States it is estimated that Sickle Cell Disease (SCD) affects 90,000 Americans. SCD occurs among 1:500 African-American births and 1:36,000 Hispanic-American births.⁴ Most infants with SCD born in the United States are now identified by routine neonatal screening. Forty-four states along with the District of Columbia, Puerto Rico and the Virgin Islands currently provide universal neonatal screening for SCD.⁵ The Sickle Cell trait occurs among about 1:12 African-Americans and 1:100 Hispanic-Americans.⁶ It is estimated that 2.5 million Americans are heterozygous carriers for the sickle cell trait.⁷

The highest prevalence of sickle cell disease is in the Eastern province where approximately 17% of the population carries the gene and 1.2% has sickle cell disease.⁸ Sickle cell disease is prevalent in many parts of India, where the prevalence has ranged from 9.4 to 22.2% in endemic areas.⁹
G6PD deficiency is one of the most common enzyme deficiency states in the world, and one of the most frequent inherited disorders. More than 400 million people (5%) are affected worldwide. The highest prevalence is in people of African, Mediterranean, and Asian heritage. The prevalence in Kurdish Jews is 50%-70%. In the US, the prevalence is approximately 10% among black males. As with all sex-linked conditions, the prevalence among females is higher but they are generally asymptomatic. In common with the carrier state of many other inherited red cell disorders (e.g., haemoglobinopathies), the deficiency of G6PD confers a degree of protection against malaria. The protection extends predominantly to hemizygous-deficient males and homozygous-deficient females.

Screening in the general population is impractical in low-risk countries. The World Health Organization does recommend screening in countries where prevalence is >3% to 5% in males. Screening of high-risk ethnic groups such as the Kurdish Jews, in whom the prevalence of G6PD is 50%-70%, and malaria endemic areas where there is a significant geographical overlap, may be worthwhile.

MATERIALS AND METHODS
This was a case-control study conducted to assess the G6PD level among Sudanese patients with sickle cell anemia. 100 samples were included in this study (50 patients who were diagnosed to have sickle cell anemia and 50 healthy subjects as controls). All patients were in stable clinical condition at time of enrollment in this study. Exclusion criteria were pregnant, diabetes, and acute infectious or chronic inflammatory disease. None of the patients were taking antioxidants or any medications that might affect the G6PD level. Blood samples were obtained from all subjects in EDTA. G6PD assay was performed at the National Reference Laboratory and Ebnaoof Hospital from July to August 2017 by full automation (Mindary BS 480). Haematological values were measured by Automated cell counter (sysmex KX-21N). This study was approved by ethical committee of the faculty of medical laboratory sciences, Alneelain University, and informed consent was obtained from each participant before sample collection and Ethical conduct was maintained during data collection and throughout the research process. All statistical analyses were performed by SPSS software version.

Continuous variables were expressed as mean and standard deviation, p-value < 0.05 was considered significant.

RESULTS

This study included 100 participants (50 cases and 50 controls). 54% were males, and 46% of the cases were females. Figure (1) represent the mean of G6PD level, it was higher (1831.8) among controls than (1808.6) in cases, P-value showed no significant difference (P= .864) between case and control group. There was significant association between low G6PD level among cases (less than 1300 - N=10) and with RBCs, Hb and PCV (Table 1).

DISCUSSION
This study utilized a quantitative approach for the determination of G6PD level among sickle cell anemia patients in Khartoum, Sudan. It included 50 Sudanese anemic Patients, their G6PD levels measured and compared with 50 age and sex matched normal subjects as controls.

We observed that 54% of the patients was males with higher G6PD activity than 46% females, this increase was insignificant according to the gender when (P-value = 0.816). Although, G6PD deficiency were determined as an X-linked recessive trait, which it is predominantly a male’s disease, present study have a similarity with Rai and Kumar, they revealed that; the percentage of G6PD deficiency in females and males is found to be 9.3% and 10.7%, respectively, so, the percentage of female deficient is quite high where the reason may be referred to the small sample size.

In addition, G6PD level among cases was lower than controls, but, no significant association (P-value = 0.864)
of low G6PD activity with anemic patients when compared with controls, this may be referred to the sample size.

Strong positive correlation was clearly appeared between RBCs, Hb, and PCV with G6PD among lower G6PD cases \((R=0.801^{*}, P=0.005)\), \((R=0.741^{*}, P=0.014)\) and \((R=0.757^{*}, P=0.011)\) this correlation could be referred to the significant changes which occurs in the structure of the outer layer of red blood cells where RBCs can break apart more readily, causing a decrease in the number of RBCs. When the body cannot produce sufficient RBCs to replace those destroyed, hemolytic anemia results and the individual may develop jaundice, weakness, fatigue, and/or shortness of breath.\(^{11}\)

**CONCLUSION**

In conclusion, low G6PD level was found in sickle cell patients, the relationship of G6PD level was not significant when compared with the controls and positively correlated with RBCs, Hb, and PCV.

**REFERENCES**