

GLYCOSYLATED HAEMOGLOBIN LEVEL IN SAUDI CHILDREN WITH SICKLE CELL ANAEMIA

Hussain A Abu Srair, FRCP(C), FAAP* Ibrahim A Al-Dabbous, CABP*
Zaki A Nasserallah, CABP* Hussain A Aman, MD*

Objective: To evaluate the glucose-6-phosphate dehydrogenase (G6PD) status in ameliorating the disease in sickle cell paediatric patient.

Design: Prospective study of patient with sickle cell anaemia and without G6PD deficiency.

Setting: Paediatric Department, Qatif Central Hospital, Saudi Arabia.

Subject: Sickle cell paediatric patient.

Results: The mean HbA_{1c} in patient with SS disease was 6.50.9 %, while that in normal paediatric patient was 6.31.5 % (P value > 0.6). On the other hand the HbA_{1c} in patient with SS disease and G6PD deficiency was 6.91.08 % which is statistically different from patient with SS disease and normal G6PD status (5.6 1.08 %) (P value < 0.04).

Conclusion: G6PD deficiency may be beneficial in reducing the haemolytic crises in Saudi paediatric SS patient.

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Sickle cell disease is a chronic haemolytic disorder, manifested by a wide variety of clinical haematological features. The prevalence of sickle cell anaemia in Saudi Arabia is well recognised^{1,2}. The incidence of the disease in the Eastern part of Saudi Arabia is particularly high³. The natural history of the disease has been reported to show mild clinical manifestation with moderate haemolytic anaemia^{4,5}.

The glycosylated haemoglobin (HbA_{1c}) level has been used as an index for the survival time of the red cells in sickle cell anaemia and other related Hb disorders^{6,7}. We have done this study to evaluate the influence of G6PD deficiency on reducing the haemolytic crises in patient with sickle cell anaemia using the HbA_{1c} as an indicator for the rate of haemolysis. .

* Paediatrics Department
Qatif Central Hospital
Tarut 31911, Saudi Arabia

METHODS

Patients attending sickle cell clinic at Qatif Central Hospital, Saudi Arabia (in their steady state), were investigated for the level of glycosylated HbA_{1c}, as well as their status of G6PD. Blood was extracted in EDTA. The glycosylated HbA_{1c} level was measured using Haleena-Glyco-Tek affinity column method, using a developer pH 8.1-8.6 containing 0.05M magnesium chloride and 0.2M glycine and glycated (Hb) eluted later using Sorbitol buffer at pH 6.0.

G6PD status was determined using commercially available kits (Boehringer Mannheim GmbH Diagnostical West Germany) where 100ul of the reagent mixed with 10ul of whole blood for 10 minutes. 10ul of the mixture put on a filter paper for 30 minutes and then read under ultra-violet lamp in a dark room. Blood with G6PD deficiency showed dark fluorescence.

All the data were entered into the computer using Stat programme. P values were done using student T test.

RESULTS

The mean value (SD) for HbA_{1c} in the 50 patients with sickle cell anaemia was 6.50.9%. This value nevertheless is not statistically different from the normal children with a mean glycosylated Hb of 6.31.5% (P > 0.6). On the other hand, the HbA_{1c} in the patient with sickle cell anaemia and G6PD deficiency (6.90.96%) was statistically different (P < 0.04) from that of patient with sickle cell anaemia and normal G6PD status (5.61.08%). Table 1 shows the data of patients with sickle cell anaemia (SSD), sickle cell anaemia and G6PD deficiency and normal children (control).

Table 1

The mean values and SD of glycosylated Hb in patients with sickle cell disease, sickle cell disease and G6PD reduced activity and control group

	Number	Mean GHbSD	P-value
Sickle cell anaemia (Total)	50	6.50.6<	%0.9 [*]
Sickle cell patient with normal G6PD	18	5.60.04>	1.08 [*]
Sickle cell anaemia with G6PD deficiency	32	6.90.96 [*]	
Normal (control)	23	6.31.51 [*]	

DISCUSSION

HbA_{1c} is due to glycosylation of the protein that occurs in a non enzymatic way throughout the life span of the rbc⁸. The level of the glycosylated Hb will therefore be influenced by the time average blood glucose concentration and the life span of the erythrocytes. Studies have shown that patients with shortened RBC life span due to haemolysis have decreased level of HbA_{1c}⁵. Based on this view we conducted our study, to see whether G6PD deficiency has any interaction with HbSS in Saudi patients.

In a previous report by Al Alayash⁹ et al, it has been shown that patients with sickle cell disease and G6PD deficiency have significantly high HbA_{1c} compared to those patients with sickle cell disease and normal G6PD status. This is in agreement with our study, but different from the observation by Al-Ali and co-workers in which both groups were found to have comparable results of HbA_{1c}¹⁰. This may be due to the fact that our sample was small, and that other in vivo factors which may affect the level of HbA_{1c} such as iron deficiency anaemia¹¹ and foetal haemoglobin level⁷ have not been considered in this study.

A possible mechanism for this is that G6PD deficiency leads to elevation of the methemoglobin level¹². This will lead to an increase in gelation time for the HbSS within the same cell. So sickling within the peripheral circulation would be averted leading to an increase in RBC life span and hence increase in HbA_{1c}. It has also been suggested that G6PD deficiency leads to an elevation of methemoglobin level which in the presence of an increase HbF, lead to a pool of HbS which can decrease deoxy-HbS solubility because of their extended volume effect¹³ and hence increase in the survival time of the RBC.

CONCLUSION

So we would like to conclude that G6PD deficiency in Saudi Sickles has a beneficial effect, in a way ameliorating the course of the disease as indexed by the elevated HbA,C.

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