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Original Research Article

Influence of glucose-6 phosphate dehydrogenase (G6-PD) deficiency upon clinico-haematological and biochemical expression of patients with sickle cell disease

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ABSTRACT

Background: According to world health organization, G6PD deficiency constitute 7.5% of world population as carriers whereas 2.9% as deficient. It has been reported from India that the prevalence varies from 0-27% in different caste, ethnic and linguistic groups. Various studies have revealed strong interaction between G6PD deficiency and sickle cell genes on one hand and environment on the other, one may therefore expect changes in their frequencies over a period of time in the population. We studied the influence of G6PD deficiency upon the clinico-haematological and biochemical expression of sickle cell disease.
Methods: The study was carried out prospectively from September 2010 to August 2011 in the department of Pathology at our institute. The material for the present study consisted of 100 cases including 76 patients with Sickle cell disease and 24 of control group. On all the samples after screening and confirmatory tests, hematological and biochemical tests were performed and discussed along with their clinical profiles.
Results: A total of 100 patients were studied, 76 were of sickle cell disease and remaining 24 patients were of control group. Majority of sickle cell trait (AS) patients (55.6%) and control patients (54%) had mild anaemia while most of the sickle cell anemia (SS) patients (44.4%) had moderate anaemia. The 2 (100%) AS and 1 (100%) control G6PD deficient patients had moderate anemia while the 2 (100%) SS patients with G6PD deficiency had severe anemia. All the sickle cell disease and control patients with G6PD deficient had elevated serum bilirubin and in all of them only unconjugated bilirubin was raised. None of the SS patients were asymptomatic. All the G6PD deficient patients had pallor and icterus. Vaso-occlusive crisis, acute chest syndrome and infectious episode were present in one (50%) each of AS and SS patients with G6PD deficiency.
Conclusions: G6PD deficiency neither exacerbated nor mitigated the frequency of painful crisis, incidence of infection or anemic episodes in patients with sickle cell disease. Hematologically no significant differences were observed in sickle cell disease patients (AS and SS) with G6PD deficiency as compared to sickle cell disease patients without G6PD deficiency and the control.

Keywords: Anaemia, Crisis, Icterus, Pallor, Unconjugated bilirubin

INTRODUCTION

Sickle cell haemoglobinopathy and glucose-6-phosphate dehydrogenase enzyme deficiency are important genetic and public health problems in Central-Eastern part of India as well. Various studies have revealed strong interaction between G6PD deficiency and sickle cell genes on one hand and environment on the other, one may therefore expect changes in their frequencies over a period of time in the population. The present study is based on finding whether such a change has taken place in these characteristics among the people of Chhattisgarh with respect to G6PD deficiency and HbS located on different chromosomes but having the same physiological effect i.e. hemolysis.

We have therefore chosen this study to assess the interaction of G6PD deficiency with SCD in Chhattisgarh, in hope that the outcome of our study will be of some relevance so that necessary actions could be taken for the better living of such group of people.

METHODS

The present study was carried out in the Department of Pathology, Pt. J.N.M. Medical College and associated Dr. B.R. Ambedkar Memorial Hospital, Raipur Chhattisgarh. The study was spread over a period of 1 year from Sep 2010 to Aug 2011.

The material for the present study consisted of 100 cases including 76 patients with Sickle cell disease and 24 of control group. The 24 control patients selected had anaemia but found to be negative for solubility test and hemoglobin electrophoresis of which 11 cases were of malaria, 8 cases of neonatal jaundice, 2 of β-thalassemia and 3 cases of miscellaneous group.

With all standard aseptic precautions, blood was collected from anti-cubital vein using 10 ml sterile plastic syringe with 21 or 22-gauge needle from each individual, after obtaining informed/written consent. 7 ml blood was collected and distributed as follows:

- 5 ml blood was delivered into ethylene diamine tetra-acetic acid (EDTA) pilot containing 1.2 mg of the anhydrous salt per ml of blood Solubility test for sickling was performed in all the samples, followed by Hb electrophoresis, complete blood count, peripheral smear, and reticulocyte count. In the entire cases qualitative dye decolourisation test for G6PD deficiency had been performed adjusting for the haemoglobin content of the patient.
- Remaining 2 ml of blood was collected in a pilot tube without anticoagulant and was sent for biochemical analysis mainly for the estimation of serum bilirubin.

RESULTS

Of the total 36 patients of AS, pallor was the commonest presentation (91.7%) and icterus, hepatomegaly and splenomegaly were present in 2 (5.6%) of them while one (2.8%) presented with vaso-occlusive crisis, acute chest syndrome and with infectious episode. None of the SS patients were asymptomatic.

Table 1: Clinical profile of the patients with and without G6PD deficiency.

Clinical feature	Patients of AS				Patients of SS				Controls			
	Without G6PD deficiency		With G6PD deficiency		Without G6PD deficiency		With G6PD deficiency		Without G6PD deficiency		With G6PD deficiency	
	N	%	N	%	N	%	N	%	N	%	N	%
Pallor	33	91.7	02	100	36	100	02	100	12	52.2	01	100
Icterus	02	5.6	02	100	11	30.5	02	100	09	39.1	01	100
Hepatomegaly	02	5.6	02	100	09	25.0	02	100	01	4.3	-	-
Splenomegaly	02	5.6	02	100	09	25.0	02	100	02	8.7	-	-
Number of vaso-occlusive crisis	01	2.8	-	-	03	8.3	01	50	-	-	-	-
Acute chest syndrome	01	2.8	-	-	02	5.6	01	50	-	-	-	-
Frequency of infectious episodes	01	2.8	-	-	02	5.6	01	50	-	-	-	-
Asymptomatic	03	8.3	-		00	00.0	-		11	47.8		
Total	**36**	**100**	**02**	**100**	**36**	**100**	**02**	**100**	**23**	**100**	**01**	**100**

All the G6PD deficient patients had pallor and icterus, hepatosplenomegaly was observed only in the patients of sickle cell disease (AS and SS) with G6PD deficiency, while vaso-occlusive crisis, acute chest syndrome and infectious episode were present in one (50%) each of AS and SS patients with G6PD deficiency Table 1. Most of the cases of AS i.e. 20 (55.6%) had mild anaemia followed by moderate anaemia in 16 (44.4%) and the 2 (100%) AS patients with G6PD deficient had moderate anemia. While most of the SS patients i.e. 16 (44.4%) had

moderate anaemia, severe anaemia was observed in 09 (25%) and the 2 (100%) SS patients who were G6PD deficient had severe anemia. Of the control patients 13 (54%) had mild anaemia and 1 (100%) control patient with G6PD deficient had moderate anemia Table 2.

Table 2: Grading of anaemia on the basis of haemoglobin values in patients with and without G6PD deficiency.

Haemoglobin value (gm/dl)	Patients of AS				Patients of SS				Controls			
	Without G6PD deficiency		With G6PD deficiency		Without G6PD deficiency		With G6PD deficiency		Without G6PD deficiency		With G6PD deficiency	
	N	%	N	%	N	%	N	%	N	%	N	%
Lower limit of normal-10 (mild anemia)	20	55.6	-	-	11	30.56	-	-	13	56.5	-	-
7- 10 gm/dl (moderate anemia)	16	44.4	02	100	16	44.4	-	-	05	21.7	01	100
< 7 gm/dl (severe anemia)	00	00	-	-	09	25	02	100	05	21.7	-	-
Total	**36**	**100**	**02**	**100**	**36**	**100**	**02**	**100**	**23**	**100**	**01**	**100**
Mean	10.2±1.89		8.85±1.33		8.5±1.16		7.21±0.57		10.7±1.91		8.2	

Of the total 36 patients of AS without G6PD deficiency most i.e. 19 (52.78%) showed microcytic and 15 (41.67%) normocytic red cells and while 2 AS patients who were G6PD deficient, 01 (50%) showed microcytic and 01 (50%) normocytic red cells. Of the SS patients 15 (41.67%) had normocytic, 10 (27.78%) had microcytic and 11 (30.56%) had macrocytic red cells, while among G6PD deficient SS patients 01 (50%) showed microcytic and 01 (50%) normocytic red cells. Of the 24 controls observed 13 (56.5%) had microcytic and 5 (21.7%) had normocytic red cells and 01 (100%) G6PD deficient control patient had normocytic red cells Table 3.

Table 3: Values of MCV in the patients with and without G6PD deficiency.

MCV (fL)	Patients of AS				Patients of SS				Controls			
	Without G6PD deficiency		With G6PD deficiency		Without G6PD deficiency		With G6PD deficiency		Without G6PD deficiency		With G6PD deficiency	
	N	%	N	%	N	%	N	%	N	%	N	%
< 80 (Microcytic)	19	52.78	01	50	10	27.78	01	50	13	56.5	-	-
80 - 100 (Normocytic)	15	41.67	01	50	15	41.67	01	50	05	21.7	01	100
>100 (Macrocytic)	02	5.56	-	-	11	30.56	-	-	05	21.7	-	-
Total	**36**	**100**	**02**	**100**	**36**	**100**	**02**	**100**	**23**	**100**	**01**	**100**
Mean	83.12 ±8.2		77.31±6.52		81.56±10.51		74.35±5.23		87.9±7.96		88.2	

Of the AS patients, most i.e. 24 (66.7%) had hypochromic and 12 (34.2%) had normochromic red cells and G6PD deficient AS patients had 1 (50%) hypochromic and 1 (50%) normochromic red cells. Of the SS patients, most i.e. 29 (80.6%) had hypochromic and 7 (19.4%) had normochromic red cells and G6PD deficient SS patients had 1 (50%) hypochromic and 1 (50%) normochromic red cells. Among the control patients 16 (66.7%) had hypochromic and 7(30.4%) had normochromic red cells while G6PD deficient control patient had 1 (100%) normochromic red cells Table 4.

The patients were categorized into three groups on the basis of corrected reticulocyte count and it was observed that among AS patients without G6PD deficiency 31 (86.1%) had normal reticulocyte count (between 0.5 to 2.5%) while 5 (13.9%) patients had values between 2.5 to 4.5% while the 02 (100%) G6PD deficient AS patients had values between 2.5% to 4.5%. Of the SS patients 16 (44.4%) had normal values of corrected reticulocyte count, 18 (50%) had values between 2.5 to 4.5%, 2 (5.6%) had values above 4.5% and the 02 (100%) G6PD deficient SS patients had values between 2.51% to 4.5%.

Among control patients 21 (91.3%) had normal values and 2 (8.7%) had values between 2.5% to 4.5% while G6PD deficient control patient 01 (100%) had value of corrected reticulocyte count within normal range Table 5.

Table 4. Values of MCH in the patients with and without G6PD deficiency.

MCH (pg)	Patients of AS				Patients of SS				Controls			
	Without G6PD deficiency		With G6PD deficiency		Without G6PD deficiency		With G6PD deficiency		Without G6PD deficiency		With G6PD deficiency	
	N	%	N	%	N	%	N	%	N	%	N	%
Hypochromic (<27)	24	66.7	01	50	29	80.6	01	50	16	69.6		
Normochromic	12	33.3	01	50	07	19.4	01	50	07	30.4	01	100
Total	**36**	**100**	**02**	**100**	**36**	**100**	**02**	**100**	**23**	**100**	**01**	**100**
Mean	27.11±2.61		26.96±2.11		25.61±2.52		24.8±2.4		27.1±2.63		29.6	

Table 5: Corrected reticulocyte count in the patients with and without G6PD deficiency.

Corrected Retic. count (%)	Patients of AS				Patients of SS				Controls			
	Without G6PD deficiency		With G6PD deficiency		Without G6PD deficiency		With G6PD deficiency		Without G6PD deficiency		With G6PD deficiency	
	N	%	N	%	N	%	N	%	N	%	N	%
0.51-2.50	31	86.1			16	44.4			21	91.3	01	100
2.51-4.50	05	13.9	02	100	18	50.0	02	100	02	8.7	-	-
≥4.51	-	-			02	5.6			-	-	-	-
Total	**36**	**100**	**02**	**100**	**36**	**100**	**02**	**100**	**23**	**100**	**01**	**100**
Mean	1.79±0.98		2.95±0.40		3.35±1.57		3.95±0.45		1.51±0.68		1.9	

Table 6: Serum bilirubin values in the patients with and without G6PD deficiency.

Serum bilirubin (mg/dl)	Patients of AS				Patients of SS				Controls			
	Without G6PD deficiency		With G6PD deficiency		Without G6PD deficiency		With G6PD deficiency		Without G6PD deficiency		With G6PD deficiency	
	N	%	N	%	N	%	N	%	N	%	N	%
≤ 1 (Normal)	27	75.0	-	-	08	22.2	-	-	08	34.8	-	-
>1 (Elevated)	09	25.0	02	100	28	77.8	02	100	15	65.2	01	100
Total	**36**	**100**			**36**	**100**	**02**	**100**	**23**	**100**	**01**	**100**
Mean	1.67±0.85		2.28±0.31		2.95±1.55		3.45±0.42		1.61±0.96		1.7	

Of the AS patients serum bilirubin was found to be within normal limit in 27 (75%) cases and elevated in 9 (25%) patients while the 2 (100%) G6PD deficient AS patients had elevated serum bilirubin, among the SS patients value was found to be normal in 8 (22.2%) and elevated in 28 (77.8%) patients and the 2 (100%) G6PD deficient SS patients had elevated serum bilirubin.

Of the control patients, 8 (33.3%) had normal values of serum bilirubin and found to be elevated in 16 (66.7%) and the single G6PD deficient control patient had elevated serum bilirubin Table 6.

Of the total 36 AS patients, conjugated bilirubin was found to be within normal limit in 34 (94.4%) patients and elevated in 2 (5.6%) patients, unconjugated bilirubin was within normal limit in 27 (75%) patients and elevated in 9 (25%) patients.

Among SS patients conjugated bilirubin was found to be within normal limit in 33 (91.7%) patients and elevated in 3 (8.3%) patients, unconjugated bilirubin was normal in 8 (22.2%) patients and elevated in 28 (77.8%) patients. In control patients conjugated bilirubin was normal in 21 (91.3%) patients and elevated in 2 (8.7%) patients while unconjugated bilirubin was normal in 8 (34.8%) patients and elevated in 16 (65.2%) patients. All the G6PD deficient AS, SS and control patients had elevated unconjugated bilirubin and normal conjugated bilirubin Table 7.

Table 7: Conjugated and unconjugated bilirubin values in the patients with and without G6PD deficiency.

Bilirubin		Patients of AS				Patients of SS				Controls			
		Without G6PD deficiency		With G6PD deficiency		Without G6PD deficiency		With G6PD deficiency		Without G6PD deficiency		With G6PD deficiency	
		N	%	N	%	N	%	N	%	N	%	N	%
Conjugated	Normal	34	94.4	02	100	33	91.7	02	100	21	91.3	01	100
	Elevated	02	5.6	-	-	03	8.3	-	-	02	8.7	-	-
Unconjugated	Normal	27	75.0	-	-	08	22.2	-	-	08	34.8	-	-
	Elevated	09	25.0	02	100	28	77.8	02	100	15	65.2	01	100

Table 8: Various studies showing clinical and hematological profile of SCD patients in relation to G6PD deficiency.

Author	Clinical feature	Severity of anemia	Hemolysis	Reticulocyte	Serum bilirubin
Smits HL et al[1]	↑ pallor, ↑ jaundice	↑	-	↑	↑
Piomelli S et al[2]	-	-	-	↑	-
Bienzle U et al[3]	-	↓	-	-	-
Gibbs WN et al[4]	-	Does not worsen anemia	No increase in hemolysis	-	-
Steinberg MH et al[5]	No significant difference	Does not worsen anemia	No increase in hemolysis	No significant difference	No significant difference
Saad STO et al[6]	-	No influence	No influence	-	-
Bouanga JC et al[7]	No advantage	Lower hemoglobin	↑ hemolysis	No significant difference	-
Bernstein SC et al[8]	Sickle Hb exerts beneficial effect on G6PD def.	-	-	-	-
Mohammad AM et al[9]	Positive correlation	-	-	-	-
Awamy BH[10]	No effect	No effect	No effect	No effect	No effect
Diop S et al[11]	No difference	-	-	-	-
Bernaudin F et al[12]	-	Does not worsen anemia	No increase in hemolysis	-	-
Nouraie M et al[13]	-	Lower haemoglobin	No increase in hemolysis	-	-
Present Study	No difference	Lower haemoglobin	No increase in hemolysis	No significant difference	No significant difference

DISCUSSION

In the present series 76 patients of sickle cell disease have been studied for G6PD deficiency along with the clinical, hematological and biochemical findings and for comparison 24 patients of anemia without sickle cell disease were studied to serve as controls.

Clinical features

In the present study there was no increase in severity of clinical features (anemia, jaundice, hepato-splenomegaly, number of vaso-occlusive crisis, number of transfusions, and frequency of infectious episodes) in G6PD deficient sickle cell patients as compared to G6PD normal sickle cell patients.

These findings were consistent with those of Bienzle U et al, Gibbs WN et al, Awamy BH and Diop S et al in that an appraisal of clinical status in sickle cell disease patients with and without G6PD showed no ameliorating or harmful effect of this enzyme abnormality upon the sickle cell disease.[3,4,10,11]

Steinberg MH et al and Bouanga et al also demonstrated that the prevalence of G6PD deficiency did not change significantly when age was stratified by decade, suggesting little survival advantage or disadvantage of the combination of G6PD deficiency and HbSS and the incidence of painful episodes, sepsis, or acute anemic episodes was similar in both groups.[5,7]

Burchard GD et al found that the spleen rates and sizes did not differ significantly between HbAA, HbAS and HbAC positive individuals of malaria endemic area. Furthermore, enlargement of spleens was found at similar

frequencies in persons with and without G6PD deficiency.[14]

Bernaudin F et al confirmed the protective effect of alpha-thalassemia and showed for the first time that G6PD deficiency and hemolysis independently increase the risk of cerebral vasculopathy in patients with sickle cell disease.[12]

Awah and Uzoegwu found that inheriting both genetic defects i.e. sickle heterozygous status and G6PD deficiency reduces the profligacy of malaria parasite and hence, ameliorates the severity of acute falciparum malaria.[15] Consequently, selective advantage against fatal falciparum malaria seems to be conferred since malarial anemia, parasitemia and severe malarial symptoms were significantly reduced. The polymorphism of haemoglobin variants and G6PD deficiency is advantageous to the community against the lethal effects of malaria especially against infection of *Plasmodium falciparum* at population level, but their combination is harmful at individual level because of low levels of red cell indices to cope with the routine human physiology. These findings get further support from the observation of Bouanga et al.[7]

Moreover, the blind administration of antimalarial drugs in such subjects will further exaggerate the normal physiology of an individual. Sometimes, it may be fatal also. Therefore, the susceptible and vulnerable communities should be screened for these genetic markers before mass administration of antimalarial (oxidant) drugs in malaria endemic localities or regions. It may be concluded that sickle cell disease and G6PD deficiency are a fairly common cause and precipitating factors for medicine induced haemolytic anemia. Early detection of these genetic disorders and avoiding indiscriminate use of precipitating medicines can prevent drug induced complications in vulnerable people.

Severity of anemia

In the present study it was observed that the mean haemoglobin level was lower in sickle cell trait (mean Hb 8.85) and homozygous sickle cell anemia (mean Hb 6.71) with co-existing G6PD deficiency in comparison with G6PD non-deficient sickle cell trait (mean Hb 10.2), G6PD non-deficient sickle cell anemia (mean Hb 8.53) and the controls patients (mean Hb 10.7). This decrease in the mean haemoglobin level was more in homozygous sickle cell patients with co-existing G6PD deficiency in comparison with counterpart sickle cell trait and controls. These measurements can reflect the intensity of hemolysis in sickle cell patients. In the present study these laboratory measurements were made while the patients were in baseline condition and not in the midst of any acute events. It therefore remains possible that Hb S patients with G6PD deficiency may be prone to the development of accelerated hemolysis when exposed to certain drugs or environmental influences that appear to provoke hemolysis in non-hemoglobinopathic patients with G6PD deficiency. This would be reflected in an increased incidence of acute anemic episodes in G6PD deficient groups. We did not observe any differences in acute anemic episodes in these patients.

In consistent with the present study no significant differences in haemoglobin values between HbAA and HbAS individuals and between G6PD deficient and G6PD normal individuals was observed in studies by Gibbs WN et al, Nieuwenhuis F et al, Steinberg MH et al, Saad STO et al and Awamy BH suggesting that there is no advantage of the association of G6PD deficiency with sickle cell disease.[4-6,10,16]

However the findings in the present study were contradictory to those of Carson and Frischer, Smits HL et al and Bouanga et al, in that a young red cell population associated with the sickle cell gene leading to elevated G6PD levels in G6PD deficient males suggests that sickle haemoglobin may exert a beneficial effect on G6PD deficiency, rather than the reverse.[1,7,17] These red cells may be better able to deal with oxidative stress (infection alone, drug alone, or a combination of both), which can precipitate severe haemolytic disease in G6PD deficiency.

However, some studies have reported that the decrease in haemoglobin is not due to increase in hemolysis. Diggs LW reviewed 747 cases of which 166 different patients with sickle cell disease were admitted in “crisis”, and observed no change in hemoglobin levels with a diminution in circulating red cells, and demonstrated that this decrease could be attributed to temporary marrow aplasia rather than to increased hemolysis.[18] Similarly Nouraie M et al reported the decrease in haemoglobin in sickle cell patients with associated G6PD deficiency is by a mechanism other than increased hemolysis.[13]

In the study by Paglialunga et al it was mentioned that G6PD plays an important role in erythroid maturation and with complete G6PD deficiency, erythroid precursors are unable to cope with high concentrations of oxygen radicals as haemoglobin synthesis is initiated, and they die by apoptosis.[19] Thus based on these findings, studies of apoptotic markers in early G6PD deficient erythroid progenitors exposed to oxidant stress and hypoxia may be beneficial.

Absolute MCV and MCH

In the present study it was observed that the mean MCV and MCH was lower in sickle cell trait (mean MCV 77.31, MCH 26.96) and homozygous sickle cell anemia (mean MCV 74.35, MCH 24.8) with co-existing G6PD deficiency in comparison with G6PD non-deficient sickle cell trait (mean MCV 83.12, MCH 27.11), G6PD non-deficient sickle cell anemia (mean MCV 81.56, MCH 25.61) and the controls patients (mean MCV 87.9, MCH 27.1). This decrease in the mean indices was more in homozygous sickle cell patients with co-existing G6PD

deficiency in comparison with counterpart sickle cell trait and controls.

These findings were consistent with those of Steinberg MH et al and Bounga et al in that the mean corpuscular volume and mean corpuscular hemoglobin were not modified by the G6PD genotypes.[5,7]

Reticulocyte count

In the present study it was observed that the mean reticulocyte count was higher in sickle cell trait (mean reticulocyte count 2.95) and homozygous sickle cell anemia (mean reticulocyte count 3.95) with co-existing G6PD deficiency in comparison with G6PD non-deficient sickle cell trait (mean reticulocyte count 1.79), G6PD non-deficient sickle cell anemia (mean reticulocyte count 3.35) and the controls (mean reticulocyte count 1.51). This increase in the mean reticulocyte count was more in homozygous sickle cell patients with co-existing G6PD deficiency in comparison with counterpart sickle cell trait and controls. These findings reflect the intensity of hemolysis in sickle cell patients. Similar findings were observed by Nouraie M et al where the G6PD deficiency was not associated with increased hemolysis as measured by reticulocyte count or a haemolytic component derived from it.[13] Gibbs W.N. et al, Steinberg MH et al and Saad STO et al too did not find any statistically significant difference in the reticulocyte count in sickle cell patients with and without G6PD deficiency.[4-6]

Smits HL et al demonstrated a brisk reticulocytosis in patients with sickle cell disease and G6PD deficiency which was due to concurrent infection or administration of certain drugs known to cause hemolysis in G6PD deficient patients.[1]

Serum bilirubin

In the present study it was observed that the mean serum bilirubin was higher in sickle cell trait (mean serum bilirubin 2.28) and homozygous sickle cell anemia (mean serum bilirubin 3.45) with co-existing G6PD deficiency in comparison with G6PD non-deficient sickle cell trait (mean serum bilirubin 1.67), G6PD non-deficient sickle cell anemia (mean serum bilirubin 2.95) and the controls patients (mean serum bilirubin 1.41). Thus no significant difference was found in the bilirubin levels in sickle cell patients with and without G6PD deficiency. It was consistent with the findings of Gibbs WN et al, Steinberg MH et al and Talafih K et al.[4,5,20] Nouraie M et al observed that the G6PD deficiency was not associated with increased hemolysis as measured by lactate dehydrogenase, serum bilirubin, aspartate amino transferase or a haemolytic component derived from these markers ($P > 0.09$) and concluded that G6PD deficiency may be associated with lower hemoglobin concentration in sickle cell anemia by a mechanism other than increased hemolysis which was consistent with the present study.[13]

However, Smits HL et al demonstrated increased jaundice as a result of accelerated hemolysis.[1]

CONCLUSION

An appraisal of clinical status in sickle cell disease patients with and without G6PD showed no ameliorating or harmful effect of this enzyme abnormality upon the sickle cell disease and it was concluded that G6PD deficiency neither exacerbated nor mitigated the frequency of painful crisis, incidence of infection or anemic episodes in patients with sickle cell disease. Hematologically no significant differences were observed in haemoglobin levels, MCV, MCH, and reticulocyte counts in sickle cell disease patients (AS and SS) with G6PD deficiency as compared to sickle cell disease patients without G6PD deficiency and the control.

However, that G6PD deficiency should be looked for in all subjects with sickle cell anemia. Since the individual with SS hemoglobin is uniquely unfit to tolerate increased hemolysis and when the two problems coexist, particular care should be exercised in the administration of drugs known to initiate hemolysis in patients with G6PD deficiency.

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