



## THE PREVALENCE OF SICKLE CELL ANEMIA AMONG PATIENTS ATTENDED PRIMARY HEALTH CARE CENTERS IN JAZAN REGION, SAUDI ARABIA

Almuhalb Gaffer Mohamed Ismail\* and Hussein Ismail Ahmed

King Saud Street Ahadalmasserha, Jazan, Saudi Arabia.

Article Received on  
02 May 2018,

Revised on 22 May 2018,  
Accepted on 12 June 2018

DOI: 10.20959/wjpps20187-11942

### \*Corresponding Author

Almuhalb Gaffer

Mohamed Ismail

King Saud Street

Ahadalmasserha, Jazan,

Saudi Arabia.

### ABSTRACT

This was an observational, descriptive, cross sectional, primary health care facility based study in Jazan province in Kingdom Saudi Arabia. The total study population of 296 sickle cell anemia patients, homozygous for HbS, who attended primary health care centers (PHCCs) in Jazan region, were investigated. Data was collected from 15 (PHCCs) randomly selected from all sectors of Jazan. The socio-demographic characteristics of the study group revealed that most of the patients in the study group (80%) were from 0-30 years old. Almost half of SCA patients in the study group (52.4%) diagnosed with early age less than a year, and 58% their general condition unwell, while 27% of them had severe disease, and 86% of them had history of

admission in hospitals the majority of them admitted in hospital twice and more. The study found that about two thirds (64%) of the patients had a history of blood transfusions. Fifth (21%) were transfused blood five times and more, 5% were transfused blood four times, and 10% were transfused blood three-times. Other associated chronic diseases were found in 22% of participants. Pain was common presenting symptom (97%), followed by fever (63%) and shortness of breath (SOB) (38%). Moreover, it was found that there was a statistically significant correlation between pain and hospitalization (P 0.006). Regarding clinical signs 80% of the patients were pale, 56% had yellow sclera and hepatomegaly found in 37%.

**INDEX:** Sickle Cell Anemia, HbS, blood transfusion, Primary Health care Centers.

## I. INTRODUCTION

Sickle cell disease (SCD) is a multisystem comprises a group of inherited blood disorders that are life-long and affect many people globally.<sup>[1]</sup>

The disease represents a major public health problem because of its associated morbidity and mortality.<sup>[2]</sup> Complications such as severe pain episodes and acute chest syndrome which need multiple hospital admissions characterized SCD patients' sufferings.<sup>[3]</sup> It is documented that SCD is associated with lower systemic blood pressures than controls and with a lower prevalence of systemic hypertension.<sup>[4,5]</sup> The disease is also associated with episodes of acute illness and progressive organ damage.<sup>[2]</sup> The higher rates of readmission of children with SCD were relatively higher than with other chronic conditions, including asthma and seizure disorder as documented by.<sup>[6]</sup>

The prevalence of the sickle cell gene in the adult population in Saudi Arabia was estimated by The Saudi Premarital Screening Program at 4.2% for sickle-cell trait and 0.26% for SCA, with the highest prevalence noted in the Eastern province (approximately 17% for sickle-cell trait and 1.2% for SCA), whereas Jazan province represents the second high prevalence (approximately 11% for sickle-cell trait and 1% for SCA).<sup>[7]</sup> The objective of this study was to measure the prevalence of SCA among patients attended primary health care centers.

## II. PATIENTS AND METHODS

### Study design

This was an observational, descriptive, cross sectional, primary health care facility based study in Jazan province in Kingdom Saudi Arabia.

### Study population

The study population included sickle cell anemia patients homozygous for HbS, who attending primary health centers in Jazan.

### Inclusion and exclusion criteria

All age groups SCA anemia patients, whom diagnoses confirmed by hemoglobin electrophoresis test, attending primary health center in Jazan were included. Individuals with Sickle Cell trait, other hemoglobinopathy, and other types of hemolytic anemia, were excluded from the study.

**Sampling procedure****Sample size**

The sample size calculated according to the equation:<sup>[8]</sup>

$$n = \frac{Nz^2pq}{(N - 1)d^2 + z^2pq}$$

n = is the sample size

N= total number of patients + primary care physicians + PHC centers.

Patients number = 3525; primary care physicians= 487; PHC centers= 173

z = z score (confidence level) = 1.96

p = level of significance = 50%

q= error = 50%

d = .05%

$$n = \frac{4185 \times 1.96^2 \times 0.5 \times 0.5}{(4185 - 1)0.0025 + 1.96^2 \times 0.5 \times 0.5}$$

n = 352

For PHCCs=  $173 \times 352 / 4185 = 15$

For primary care doctors =  $487 \times 352 / 4185 = 41$

For SCA patients =  $3525 \times 352 / 4185 = 296$

**Sampling technique**

If the PHCC revisited again by the investigator till the adequate numbers reached. Each patient was registered and interviewed on the clinic by the investigator using a pre-tested and pre-coded questionnaire.

**Tool of data collection**

SCA patients were monitored for history and examination. The patients were registered and interviewed in the clinic by the investigator using a pre-tested and pre-coded questionnaire.

**Data processing and analysis**

All data collected were coded for subsequent computer processing and analysis on a personal computer using SPSS program. Chi-squared test was used to assess the level of significance of the differences between proportions.

### Ethical consideration

The study was approved from Research Ethics Committee Jazan General Hospital. Written consent was taken from the patients or their parents after clarifying the objectives and process of study.

### III. RESULTS

The socio-demographic characteristics of the study group are shown in Fig 1. From the total number (296) of patients who were diagnosed as having sickle cell anemia, 172 (58%) of them were female, and 124 (42%) were male. Patients in the age group 0-5 years constituted 9%, age group 6-15 years 28%, age group 16-30 years 43% and age group 31-45 year, and more than 46 year were 16% and 4%, respectively.

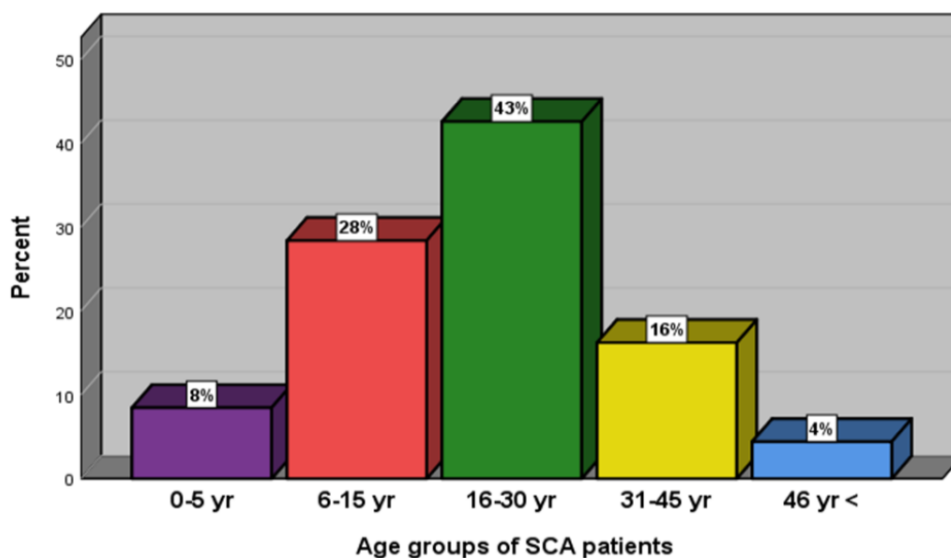
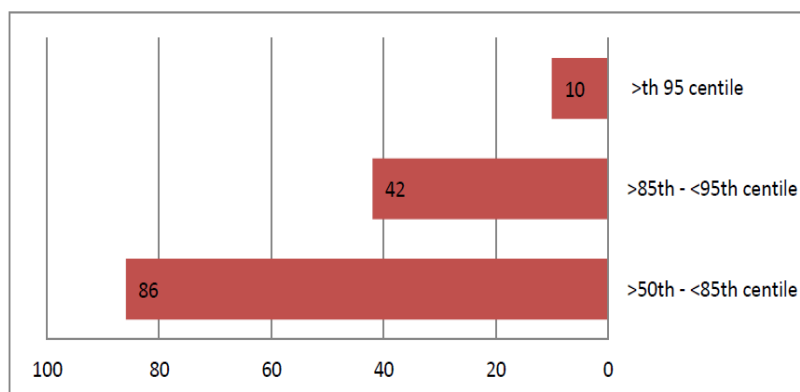


Figure 1: Age characteristics of the study group.

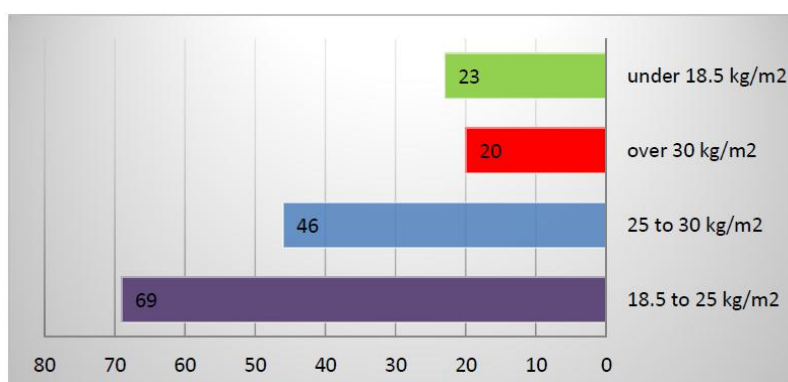
### Clinical information of the study group

A total number of 138 SCA patients less than 19 years Saudi were weighed for stature percentiles used to determined obesity. It was observed that, the body mass index of 86 (62%) was >50th-<85th centile, while the body mass index of 42 (30%) of the patients was >85th-<95th centile, and 10 (8%) their body mass index >95th centile (Figure 2).



**Figure 2: Body mass index of SCA patients aged 2-19 yrs.**

In contrast, 69 (44%) of 158 SCA patients who were more than 19 years old had normal body mass index (18.5-25 kg/m<sup>2</sup>), and 46 (29%) had body mass index between 25-30 kg/m<sup>2</sup>, 23 (14%) who had body mass index below 18.5 kg/m<sup>2</sup>, where the remaining 20 (13%) had obesity with body mass index more than 30kg/m<sup>2</sup> (Figure 3).



**Figure 3: Body mass index of SCA patients aged more than 19 yrs.**

It was also found that 191 (64%) of the patients had history of blood transfusions. Sixty three patients (21%) were transfused blood five times and more, 15 (5%) were transfused blood four times, 31 patients (10%) were transfused blood three-times, 24 patients (8%) were transfused blood twice, and 58 patients (20%) were transfused blood once (Table 1).

**Table 1: Number of blood transfusions for children with sickle cell anemia.**

		Frequency	Percent
Valid	No transfusion	105	36
	five times and more	63	21
	four time	15	5
	three time	31	10
	Twice	24	8
	Once	58	20
	Total	296	100

Table 2 shows the frequency of chronic diseases in SCA patients (n=296) Twenty patients were known hypertensive, 16 with Diabetes Mellitus, 8 with asthma, 5 patients developed hepatitis B, 6 patients developed Psychiatric disease, and thalassemia coexist in 5 patients, as well as, Thyrotoxicosis, Irritable bowel syndrome, and Osteoporosis, were 4, 2,1, respectively and 65 (22%) of SCA patients had other chronic disease.

**Table 2: The frequency of chronic diseases in SCA patients.**

	Frequency	Percent
Had no chronic diseases	164	55.4
Hypertension	20	6.7
Diabetes mellitus	16	5.4
Asthma	8	2.7
Hepatitis B	5	1.7
Psychiatric disease	6	2
Thalassemia	5	1.7
Thyrotoxicosis	4	1.4
Irritable bowel syndrome	2	0.7
Osteoporosis	1	0.4
Other chronic disease	65	22
Total	296	100

The severity of the disease in patients (N= 296) is shown in Table (3). Any patient who scored  $\geq 5$  was considered as a severe case (79), those with scores of 3-4 were considered as moderate cases (103), and those with scores of 0-2 were considered as mild cases (123) (Table 3).

**Table 3: Severity of the disease in patients.**

		14/ HGB: mg/dl				Total
		<6	6-7.9	8-10	>10	
Admission:	No	0	5	20	14	39
	1-2 times	0	16	39	29	84
	3-4 times	7	21	19	7	54
	5 and more	7	65	38	9	119
Total		14	107	116	59	296
		14	107	116	59	
		severe SCA		moderate SCA		mild SCA

Disease symptoms at the time of previous visit to clinic were variable. Pain was found in 287 (97%) patients, 185 (63%) had fever, 114 (38%) had Shortness of breathing, 86 (29%) had chest pain, 63 (21.3%) had abdominal pain, 61 (21%) had failure to thrive or cachexia. Other symptoms were found in minor proportions as shown in (Table 4).

**Table 4: Common symptoms of SCA patients (N=296).**

		Frequency	Percent
Pain		287	97
Fever		185	63
Chest pain		86	29.0
Shortness of breathing		114	38
Abdominal pain		63	21.3
Loss of weight		61	21
Other	Headache	23	7.8
	Yellowish of sclera	6	2.0
	Palpitation	4	1.4
	Burning micturition	2	0.7
	Dizziness	2	0.7

Regarding complication of sickle cell anemia, 49 (16.6%) of patients had history of cholelithiasis, 17 (5.7%) with cardiac involvement, 12 (4.1%) with history of priapism, 14(4.7%) patients underwent to splenectomy, and 9 (3%) had depression, as well as 5(1.7) patients developed stroke. Other complications were found in different variables as shown in (Table 5).

**Table 5: Complications of SCA patients (N=296).**

		Frequency	Percent
Cholelithiasis		49	16.6
Cardiac involvement		17	5.7
History of priapism		12	4.1
Underwent splenectomy		14	4.7
Depression		9	3.0
Stroke		5	1.7
Other	Renal involvement	7	2.4

#### IV. DISCUSSION

In this study the socio-demographic characteristics of the study group revealed that most of the patients in the study group (80%) were from 0-30 years old. This may indicate decreased life span with age due to different complications of SCD. This finding was similar to those reported in other studies by.<sup>[9,10]</sup>

This research revealed that children and teenagers with SCA had roughly the same proportion of obesity and overweight to their peers in Saudi community. This finding was different from that reported by<sup>[11]</sup> when compared to African patients who are mostly growth retarded. The

rate of obesity revealed in this study was less by two fold from the Saudi rate of obesity according to the National Health Information Survey in 2013.<sup>[12]</sup>

Blood transfusion is recommended for certain severe SCD complications such as acute chest syndrome, splenic or hepatic sequestration.<sup>[13]</sup> In the present study the rate of patients with SCD who received simple red blood cell transfusion was similar with prior observations by [14] who reported (23 - 36%).

The rate of SCD accompanying hypertension in this study is lower than those reported by<sup>[15]</sup> who recorded that 44% had SBP of 120–139 mm Hg or DBP of 70–89 mm Hg (classified as relative hypertension) and 10% had SBP  $\geq$ 140 mm Hg or DBP  $\geq$ 90 mm Hg (classified as hypertension).

The low prevalence of diabetes mellitus in SCD patients compared to the general population may be attributed to the fact that a risk genetic variation causing a progressive disease such as diabetes may exhibit certain effects on metabolism in early life. This finding is consistent with the findings reported by.<sup>[16]</sup> Asthma is a common comorbidity in patients with sickle-cell disease, occurring in 15–28% of participants in large multicenter cohort studies.<sup>[17,18,19]</sup> This finding was more than that reported in our study.

In our study the degree of severity of SCA was assessed in participants according to HbG level and hospitalization frequency. Our study revealed that the frequency of patients with severe SCD was lower than that type described for Sudanese.<sup>[11, 20, 21]</sup> The study also recorded that the majority of patients were classified as moderate and mild disease. This may be attributed to the haplotype of Saudi in addition to other factors such as environmental factors.

Acute painful crisis continues to be the primary reason for most health care visits for patients with SCD. The results of this study presented that the majority of patients were diagnosed with painful episodes. Furthermore, painful events were significantly and positively associated with in-patient hospitalizations of SCD patients in the current study. These results were comparable to data obtained by other studies in Saudi Arabia, Middle East and Africa.<sup>[11,17,22,23]</sup> Fever was found to be the second-leading cause of visits to clinics among SCD patients in Jazan, followed by 9 chest pain then SOB. Although infections contributed to a considerable percentage of healthcare utilization events, an earlier study in Basra reported a lower rate of infections among hospitalized children with SCD compared to this study.



Likewise, this finding is in agreement with<sup>[17,24]</sup> who have cited fever as a reason for ED visits in patients with SCD. As would be anticipated, as result to the breakdown of hemoglobin in SCD patients, cholelithiasis occurred in a considerable proportion of participants. The result of this study was lower than that observed in Nigerians with SCD, which may to be affected by local diet and possible genetic factors.<sup>[25]</sup> Likewise, cardiac involvement detected in a very small number of patients of SCD in this study, was less than data obtained from other study.<sup>[11]</sup>

In the current study the frequency of patients with SCD diagnosed with a depressive disorder was lower than that previously reported by,<sup>[26,27]</sup> in either adult or adolescent samples using depression rating scales (26% and 29%, respectively).

## V. CONCLUSION AND RECOMMENDATIONS

This research concluded that children and teenagers with SCA had roughly the same proportion of obesity and overweight to their peers in Saudi community. The rate of SCD accompanying hypertension in this study was generally lower than those reported in the literature and the frequency of patients with SCD diagnosed with a depressive disorder was lower than that previously reported. The study recommended promotion of genetic counseling and management of SCD clinics or centers in areas where disease was more prevalent. Further studies shall be conducted to assess the complications of disease.

## VI. REFERENCES

1. Hussain R, Yusuf, Michele A, Lloyd-Puryear, Althea M, Grant, Christopher S, Parker, Melissa S, Creary and Hani K. Atrash Sickle Cell Disease: The Need for a Public Health Agenda. *Am J Prev Med*, 2011; 41(6S4): S376–S383.
2. R.D. Cancado, “Sickle cell disease: looking back but towards the future,” *Revista Brasileira de Hematologia e Hemoterapia*, 2012; 34(3): 175–177.
3. Sobota A, Graham DA, Neufeld EJ, Heeney MM. Thirty-day readmission rates following hospitalization for pediatric sickle cell crisis at freestanding children’s hospitals: risk factors and hospital variation. *Pediatr Blood Cancer*, 2012; 58: 61–5. [PubMed: 21674766]
4. Ernst AA, Weiss SJ, Johnson WD, Takakuwa KM. Blood pressure in acute vasoocclusive crises of sickle cell disease. *South Med J.*, 2000; 93(6): 590–2. [PubMed: 10881775]

5. Guasch A, Navarrete J, Nass K, Zayas CF. Glomerular involvement in adults with sickle cell hemoglobinopathies: prevalence and clinical correlates of progressive renal failure. *J Am Soc Nephrol.* 2006; 17: 2228–2235. [PubMed: 16837635]
6. Berry JG, Toomey SL, Zaslavsky AM, et al. Pediatric readmission prevalence and variability across hospitals. *JAMA,* 2013; 309(4): 372–380.  
<http://dx.doi.org/10.1001/jama.2012.188351>.
7. Wasil Jastaniah 2011. Epidemiology of sickle cell disease in Saudi Arabia by in, 2011. May-Jun <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3119971>.
8. Lwange SK, Lemeshow S. Sample size determination in health studies. *Pract Manual,* 1987; 25-6.
9. Fk HF, Hood WE. The heart in sickle cell anaemia. *ArchIntern Med,* 1982; 142: 1680-684.
10. Platt O, Nathan DG. Sickle cell disease. In: Nathan D G, Oski FA, (editors). *Haematology of Infancy and Childhood.* 2nded .Philadelphia, WB Saunders Company, 1989; 617-625.
11. Ghada Osman Mohamed Ali. Cardiac Manifestations of Sickle Cell Anaemia in Children in Khartoum. A Thesis Submitted in Partial Fulfilment for the Requirement of the Clinical MD Degree in Paediatric and Child Health. April, 2004.
12. obesity control program- MOH-  
KSA<https://www.moh.gov.sa/OCP/Pages/DirectorMessage.aspx>.
13. Yawn BP, Buchanan GR, Afeniyi-Annan AN, Ballas SK, Hassell KL, James AH, Jordan L, Lanzkron SM, Lottenberg R, Savage WJ, Tanabe PJ, Ware RE, Murad MH, Goldsmith JC, Ortiz E, Fulwood R, Horton A, John-Sowah J. Management of sickle cell disease: summary of the 2014 evidence-based report by expert panel members. *JAMA,* 2014; 312: 1033–48. [PubMed: 25203083].
14. Mehdi Nouraie and Victor R. Gordeuk. Blood transfusion and 30-day readmission rate in adult patients hospitalized with sickle cell disease crisis. USA, 2015.
15. Victor R. Gordeuk, MD, Vandana Sachdev, MD, James Taylor, MD, Mark T. Gladwin, MD, Gregory Kato, MD, and Oswaldo L. Castro, MD. Relative Systemic Hypertension in Patients with Sickle Cell Disease is Associated with Risk of Pulmonary Hypertension and Renal Insufficiency Howard University, USA, 2015.
16. Xu Zhang, Wei Zhang, Santosh L. Saraf, Mehdi Nouraie, Jin Han, Michel Gowhari, Johara Hassan, Galina Miasnikova, Adelina Sergueeva, Sergei Nekhai, Rick Kittles,7 Roberto F. Machado, Joe G N Garcia, Mark T. Gladwin, Martin H. Steinberg, Paola

- Sebastiani, Donald A. McClain, and Victor R. Gordeuk. Genetic polymorphism of APOB is associated with diabetes mellitus in sickle cell disease, 2015.
17. Zeina A. Salman and Meaad K. Hassan. Hospitalization Events among Children and Adolescents with Sickle Cell Disease in Basra, Iraq, 2015.
  18. An P, Barron-Casella EA, Strunk RC, Hamilton RG, Casella JF, DeBaun MR. Elevation of IgE in children with sickle cell disease is associated with doctor diagnosis of asthma and increased morbidity *J Allergy Clin Immunol*, 2011; 127: 1440–46. [PMC free article] [PubMed].
  19. Strunk RC, Cohen RT, Cooper BP, et al. Wheezing symptoms and parental asthma are associated with a physician diagnosis of asthma in children with sickle cell anemia. *J Pediatr*, 2014; 164: 821–26. e1.[PMC free article] [PubMed].
  20. Ahmed HA, Baker EA. Sickling in the Sudan; Result of survey in Blue Nile province. *East Afr Med J.*, 1986; 6: 395-99.
  21. Archbald RG. A case of sickle cell anaemia in the Sudan. *Trans R Soc Trop Med Hyg.*, 1926; 19: 389-93.
  22. Paula Tanabe, Christopher Reddin, Victoria L. Thornton, Knox H. Todd, Ted Wun, and John S. Lyons. Emergency Department Sickle Cell Assessment of Needs and Strengths (ED-SCANS), a Focus Group and Decision Support Tool Development Project. *Acad Emerg Med.*, 2010; 17(8): 848–858.
  23. Jaiyesimi, F., Pandey, R., Bux, D., Sreekrishna, Y., Zaki, F., Krishnamoorthy, N. Sickle cell morbidity profile in Omani children Nazowa, Oman, 2002.
  24. Koshy M, Leikin J, Dorn L, Lebby T, Talischy N, Telfert MC. Evaluation and management of sickle cell disease in the emergency department (AN 18-YEAR EXPERIENCE): 1974-1992. *Am J Ther*, 1994; 1(4): 309–20. doi: 10.1097/00045391-199412000-00011. [PubMed] [Cross Ref].
  25. Durosinmi MA1, Ogunseyinde AO, Olatunji PO, Esan GJ. Prevalence of cholelithiasis in Nigerians with sickle cell disease, 1989.
  26. Jeanette M. Jerrell, Avnish Tripathi, and Roger S. McIntyre. Prevalence and Treatment of Depression in Children and Adolescents With Sickle Cell Disease: A Retrospective Cohort Study, South Carolina Medicaid medical and pharmacy claims between January 1, 1996, and December 31, 2006.
  27. Monika R Asnani, Raphael Fraser, Norma A Lewis, and Marvin E Reid. Depression and loneliness in Jamaicans with sickle cell disease. *BMC Psychiatry*, 2010.