

Clinical Presentation of Sickle Cell Disease in Patients Admitted To Al Obied Specialized Hospital-Al Obied-Sudan

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Received: 01 Mar 2022; **Accepted:** 03 Apr 2022; **Published:** 08 Apr 2022

Citation: Toro SA, Mohamed Al Mustafa AZ, Bazie EA. Clinical Presentation of Sickle Cell Disease in Patients Admitted To Al Obied Specialized Hospital-Al Obied-Sudan. *J Med - Clin Res & Rev.* 2022; 6(4): 1-5.

Keywords

Sickle Cell disease, Blood transfusion, Transcranial doppler imaging, Hydroxyurea.

Introduction

Sickle Cell disease is an increasing global health problem; it is estimated to be about 300,000 born with sickle cell anemia and expected to rise to 400,000 by 2050. Quality of life and survival of patients with sickle cell disease was dramatically improved in the last years after early diagnosis, penicillin prophylaxis, blood transfusion, transcranial doppler imaging, hydroxyurea, and hematopoietic stem-cell transplantation. [1].

Sickle cell disease is a life-threatening hematological disorder with Pain as the hallmark presentation. Pain and its complications are spared during infants' lives because of their elevated HbF levels. The first bone affected in infants is the small bones in hands and foot, called 'dactylitis.' Dactylitis developed in half of the children by two years. [3]

The sickle-cell crisis occurs from disrupted blood flow in small vessels by abnormal sickle-shaped erythrocyte that leads to vaso-occlusion that lead to distal tissue ischemia and inflammation with symptoms of painful crises. Repeated disrupted blood flow due to sickling and ongoing hemolytic anemia, even asymptomatic, lead to parenchymal injury and chronic organ damage, causing substantial morbidity and early mortality [4].

In Sudan, Ahmed and his colleagues reported prevalence of sickle cell disease was 30% [5] and 16% among immigrants from the Blue Nile province, and a separate study has done by Foy et al. reported 18% among Nilotic tribes in the south of Sudan [6].

The severity of sickle cell disease could be treated and managed by factors that can modify disease severity, such as fetal hemoglobin (HbF) levels and α -thalassemia; other genetic variants might affect specific subphenotypes. The influence of altitude and temperature on patients with sickle cell disease; has been recently studied as nongenetic factors, including climate and air quality, suggest more complex associations between environmental factors and clinical complications [7].

The management of SCD and its complications are required to maintain a good quality of life needed supportive management, symptomatic treatment, and preventative measures [8,9].

A published guideline [10] recommends daily oral prophylactic penicillin for children up to 5 years and immunization to reduce the risk of infections [11].

Objectives

General objective

To study the Clinical Presentation of Sickle Cell Disease in Patients Admitted to Al Obied Specialized Hospital-Al Obied-North Kordofan State -Sudan.

Specific objectives

- To determine the demographic data of SCD children admitted to ALObied Specialized Hospital during study period.
- To identify the clinical presentations of sickle cell disease children admitted to Al Obied Specialized Hospital.
- To find out patients on hydroxyurea and folic acid among study group.

Materials and Methods

Study design

Retrospective, cross-sectional, hospital-based study.

Study area and setting

Study conducted at Al Obied Specialized Hospital, North Kordofan State. Al Obied Specialized Hospital is the only tertiary hospital in the State; also, it covers all patients from South and West Kordofan States.

Study sample

All patients with Sickle Cell Diseases admitted to Al-obaid Specialized Hospital during study period were included.

Study duration

- The study conducted within the period from 1st January 2018 to 1st January 2019.

Study population

Patients who were admitted to Al Obied Specialized Hospital during study period.

Study inclusion criteria

All Sickle cell disease children who diagnosed by Hb electrophoresis and admitted to Al-obaid specialized hospital during study period.

Study exclusion criteria

1. Patients more than 14 years old
2. Non-sickle cell diseases patients
3. Incomplete medical records and documentations.

Data collection tools and methods

Data on demographic variables, diagnosis during admission, will be collect from patients, registration files through checklist by investigator.

Data analysis

Data entered, and analyzed using statistical package for social sciences (SPSS) version 25.0.

Bi variable analysis will be applied to assess the pattern of admission with chi square test. P value of less than 0.05 will be considered significant.

Ethical considerations

Written ethical clearance and approval for conducting this research obtained from Sudan Medical Specialization Board ethical committee.

Written consent from center administration and treating doctor, confidentiality and secrecy maintained.

Result

We studied 201 patients with sickle cell anemia who attended the ELOBIED Specialized hospital in North Kordofan State hematology clinic. Males were 113(56.2%), and Females were 88(43.8%) with a Male: Female ratio of 1.2:1 (Figure 1).

Age distribution showed 84 (41.8%) patients aged 1_5 years old, 62(30.8%) patients were aged 6_10 years old, 43(21.4%) patients were 11_14 years old, and 12(6%) patients were less than one years old (Figure 2).

The residence distribution

116(57.7%) were Urban residency. 77(38.3%) were rural residency, and 8 (4%) were nomads residency (Figure 3).

The tribe's distribution of the patients showed: 28(13.9%) patients were Bedaria, 25(12.4%) patients were Hausa, 23(11.4%) patients were Hawasma, 21(10.4%) patients were Bargo, 19(9.5%) patients were Galaba Hawara, 17(8.5%) patients were Gawamaa, 12(6%) patients were Masseriaa, 11(5.5%) patients were Barno, 11(5.5%) patients were Dar-Hamid, 8(4%) patients were Kenanaand, 26(12.9%) patients were other tribes (Figure 4).

Health Insurance was found with 163 (81.1%) of patients, and 38(18.9%). had no health insurance (Figure 5).

Family members with sickle cell anemia were found in 104 (51.7%) patients, while 97 (48.3%) patients had no family history (Figure 6).

Age of first clinical symptoms of sickle cell anemia showed: 82 (40.8%) patients were in less than six months of age, 74 (36.8%) patients in 6_12 months of age, and 45 (22.4%) patients after 12 months of age Table 1.

Our result showed 92% of patients on folic acid and 72.6% on hydroxyurea (Table 2).

In our study, Blood transfusion was done to 166 (82.6%) patients, exchange transfusion was done to 21(10.4%) patients, and 10(4.76%) were in regular blood transfusion following stroke (Table 3).

Clinical presentations during admission showed 88 (43.8%) patients had Bone pain, 21 (10.4%) patients had pneumonia, 18 (9%) patients had anemic heart failure, 14 (5.5%) patients had a stroke, 12 (7%) patients had malaria, 9 (4.5%) patients had acute chest syndrome, Five (2.5%) patients had Aplastic crisis, Four (2%) patients had UTI, and 30 (14.8%) patients had other symptoms (Figure 7).

There was a significant association between the clinical presentation of sickle cell anemia and Hydroxyurea supplementation, *P-value* =000 (Table 4).

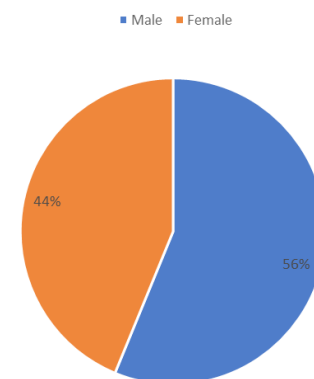


Figure 1: Gender Distribution.

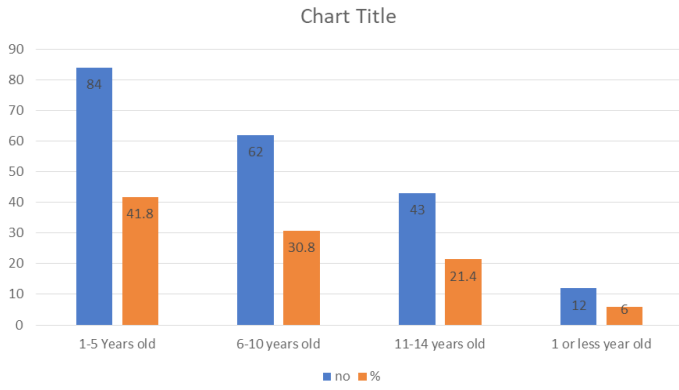


Figure 2: Age distribution.

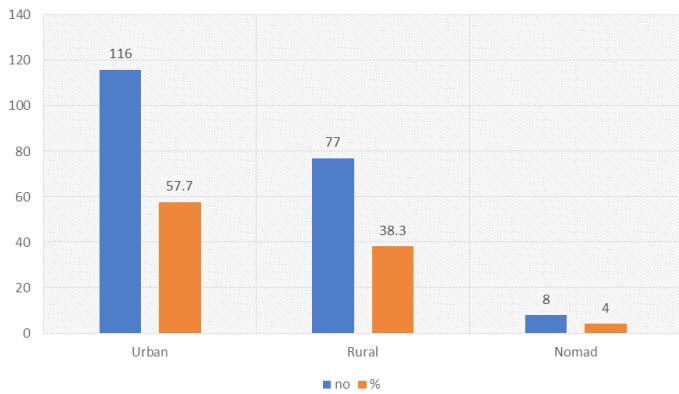


Figure 3: Residency distribution.

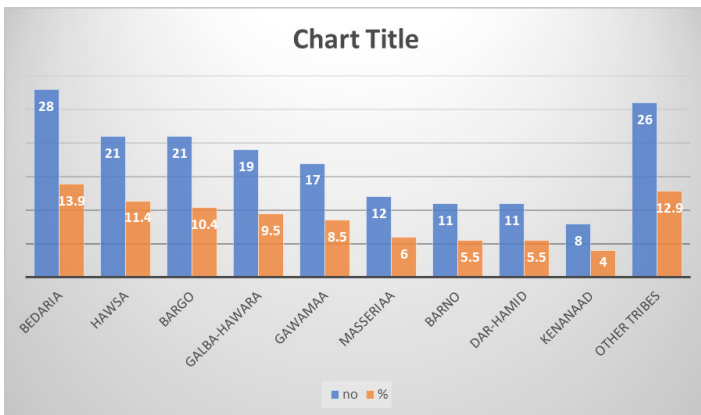


Figure 4: Tribes distributions.

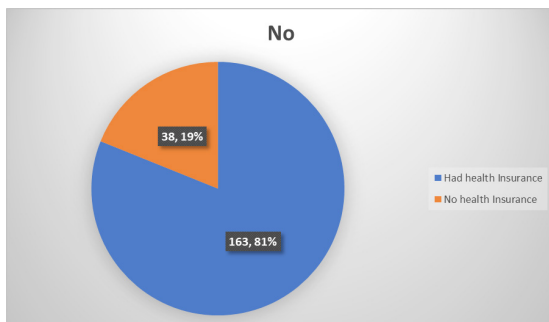


Figure 5: Health Insurance.

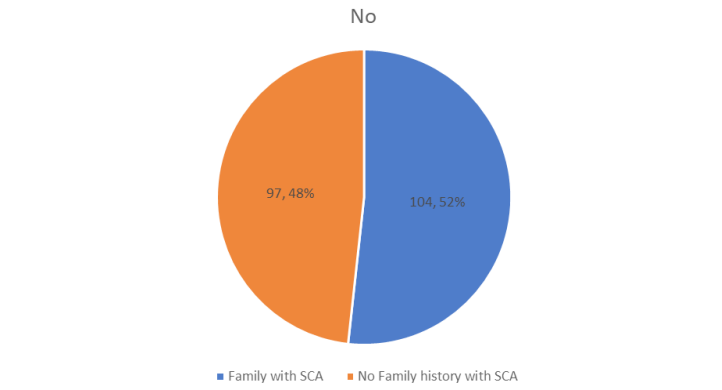


Figure 6: Family history with SCA.

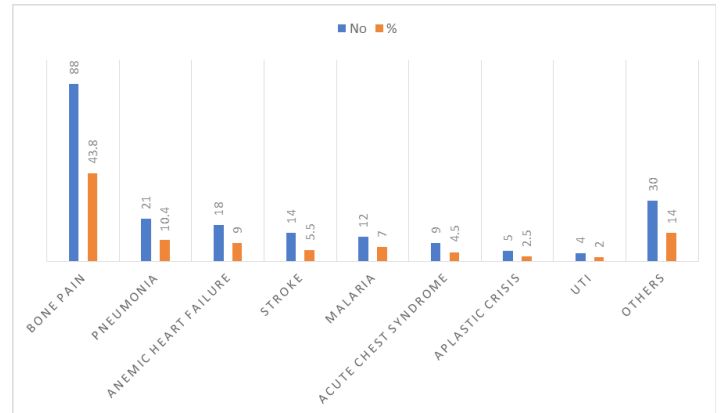


Figure 7: Clinical Presentations Distribution.

Table 1: Participants Distribution According To Age at First Diagnosis.

Age group	Frequency	Percent
Less than 6 month	82	40.8%
(6-12) month	74	36.8%
More than 12 months	45	22.4%
Total	201	100%

Table 2: Folic Acid and Hydroxyurea Use among Study Participants.

		Frequency	Percent
Folic acid complain	Yes	185	92%
	No	16	8%
Hydroxy urea complains	Yes	146	72.6%
	No	55	27.4%

Table 3: Blood Transfusion and Exchange Transfusion among Participants.

Percent	Frequency		
82.6%	166	Yes	Blood transfusion
17.4%	35	No	
10.4%	21	Yes	Exchange transfusion
89.6%	180	No	

Table 4: Clinical Presentation and Hydroxy Urea Use Crosstabulation.

Clinical presentation	Hydroxy urea complians		Total
	Yes	No	
Bone pain	48 60.8%	31 39.2%	79 100.0%
Acute chest syndrome	6 66.7%	3 33.3%	9 100.0%
A Plastic crisis	1 50.0%	0 0.0%	2 100.0%
Pneumonia	18 90.0%	2 10.0%	20 100.0%
Anemic heart failure	11 73.3%	4 26.7%	15 100.0%
Stroke	9 81.8%	2 18.2%	11 100.0%
Malaria	8 72.7%	2 18.2%	11 100.0%
UTI	3 75.0%	1 25.0%	4 100.0%
Others	24 80.0%	6 20.0%	30 100.0%
Total	128 70.7%	51 28.2%	181 100.0%

P-Value= 0.000

Discussion

Retrospective, cross-sectional, hospital-based study conducted at Al Obied Specialized Hospital, North Kordofan state, included two hundred one sickle cell disease patients, majority of the 41.8% were 1-5 years old, and only 6% were less than one years old. Within the period from 2018 to 2019.

In our study, males were predominant, with a male: female ratio of 1.2:1; Mustafa AE et al., in a study in the same hospital, reported similar findings with the same male: female ratio (1.2:1), which strongly support our study findings [12]. Muna Jawish et al. [13] in Saudi Arabia showed male predominance and most of their patients were between 5 to 10 years old.

Sickle cell anemia is reported among Kordofan and Darfur region mainly among Albaggara, an Afro-Arab constellation of tribes with predominantly African descent, in addition to Bedelia, Fulani, Messeryia, Hummer, Berge, Fur, and Masaleet [14]. In our study, the main tribes affected were Bedariah, Falatah, and Hausa, as they were the main tribes in the North Kordofan region. Mustafa AE et al. [16] study showed Falatah are the common tribe affected with sickle cell anemia in the same hospital. In Algardaref State, Osman et al. [15] reported Masalet tribe are more affected; in West Kordofan, the disease affects up to 30% of the Masseria tribe as reported by Ahmed et al. [16].

We found 51.7% of our study group had a family history of sickle cell anemia; we supposed the percentage of family members affected with sickle cell anemia might be more than this as families deny the disease as it is a social stigma in their communities.

Bone pain was reported in most of our patients, followed by pneumonia and anemic heart failure, respectively, acute chest

syndrome in 4.5%, a plastic crisis in 2.5%, stroke in 5.5%, and malaria in 7%. Mustafa AE et al. [12] reported anemia in 79%, joint swelling (35%), and bone swelling (31%). Osman et al. [15] had results near to our study as it showed hand-foot syndrome in 16% and pneumonia in 8%. Elderberry et al. also reported anemia as clinical presentation in sickle cell anemia [17]. In Northern Darfur tribes, Mustafa MH et al. [18] in Higlig South Kordofan State, and Macharia et al. [19] in Kenya, studies. In addition, Adegoke SA [96] revealed that 33.1% had a previous history of dactylitis (hand-foot syndrome). Muna Jawish et al. [13] in their study painful crises is the commonest presentation.

We found most of our patients had the first clinical symptoms of sickle cell anemia by less than six months of age. Mustafa AE et al. [12] located about 80% of the children were initially diagnosed at age less than five years, most of our patients presented with first symptoms below five years old.

The present study showed that 92% of patients used folic acid, and 72.6% used Hydroxyurea. Also, a study done in 2014 by Ruchita Dixit et al. [21] showed that it was unclear if folate supplementation has any effect on hemoglobin concentration, growth, minor infections, major infections, acute splenic sequestration, dactylitis, or episodes of bone or abdominal pain. Jose J et al. [22] showed that Hydroxyurea had significant and excellent outcomes in suitable compliance patients. Our study showed a significant association between clinical presentation and compliance to Hydroxyurea.

Blood transfusion was done on 83% of our patients, and it was nearly like Gülendarm Karadağ et al. [23] study, which was 77.2%. Muna Jawish et al. [13]. In their study, blood transfusion is only for 8.2% unlike our finding.

Conclusion

Bedaria tribes are more affected than other tribes, which their predominance can explain. Vasocclusive crises are common in our patients, and Malaria has a high prevalence.

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