

## The prevalence of leukocyte abnormalities among Sudanese patients with sickle cell disease

Tarig Osman Khalafallah Ahmed <sup>1,\*</sup>, Ekhlash Alrasheid Abu Elfadul <sup>1</sup>, Ahmed A. Agab Eldour <sup>2</sup> and Omer Ibrahim Abdallah Mohammed <sup>3</sup>

<sup>1</sup> Faculty of Medical Laboratory Science, Department of Hematology, University of Kordofan, El-Obeid, Sudan.

<sup>2</sup> Faculty of Medicine, Department of Pathology, University of Kordofan, El-Obeid, Sudan.

<sup>3</sup> Sharjah Municipality, Department of Public Health Clinic, Sharjah- United Arab Emirates.

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### Abstract

Sickle cell disease (SCD) is an inherited blood disorder that affects red blood cells. The study was conducted in Elobied town during the period May 2011 to September 2011. The aim of this study is to detect the abnormalities of leucocytes among sickle cell anemic patients. 40 sickle cell anemic patients; age range between 8 months to 23 years. Blood sample was taken for all patients and the laboratory investigation were performed using automated estimation for: hemoglobin (Hb), Packed cell volume (PCV), red cell count (RBCs), mean cell volume (MCV), mean cell hemoglobin (MCH), mean cell concentration (MCHC), and total white blood cells, comment on blood film using manual methods. The conclusion of this study there is increase in total white blood cells with shift to left in neutrophil precursor in sickle cell patients with complications, the most immature cells are band form, myelocytes and metamyelocytes, and there also lymphocytosis and neutrophilia which has been increases in response to infections.

**Keywords:** Leukocyte abnormalities ; SCD ; Sudanese patients; CBC

### 1. Introduction

Sickle cell disease is a chronic hemolytic disorder that is marked by tendency of hemoglobin molecules within red cells to polymerise and deform the red cell into a sickle (or crescent) shape resulting in characteristic vasoocclusive events and accelerated hemolysis [1,2]. White blood cell (WBC) or leukocyte count is a blood test used to detect a range of blood disorders, evaluate the severity of various underlying diseases, and at times predict the risk of death. High white blood cell (WBC) count is an indicator of sickle cell disease (SCD) severity [3]. Bacterial infection area major cause of morbidity and mortality in children with sickle-cell disease. The increased susceptibility of affected children is likely to result from several causes, including impaired splenic function, defects in complement activation, micronutrient deficiencies, and tissue ischemia [4]. Leukocytosis, in the absence of infection, is common in SCD patients and predicts for stroke, acute chest syndrome, and overall mortality. Neutrophils and monocytes have been shown to be activated in these patients [5]. Patients with impaired ability of leukocytes to kill microbes are more prone to infections, which precipitate sickle cell crisis. Reduction of leukocyte count ameliorates SCD. Similarly, targeted blockade or reduced synthesis of specific leukocyte adhesion molecules and their ligands might confer clinical benefit in SCD [6]. The pathogenesis of sickle cell vascular disease is complex and not yet fully understood [5]. The purpose of the current study was to estimate the prevalence of abnormal white blood cells number and types in a sample of Sudanese patients with SCD.

\* Corresponding author: Tarig Osman Khalafallah Ahmed  
Faculty of Medical Laboratory Science, Department of Hematology, University of Kordofan, El-Obeid, Sudan.

## 2. Material and methods

The current study is Cross-sectional study was conducted in Elobied town during the period May 2011 to September 2011. Ethical clearances were obtained from Kordofan University Ethical committee. Any patient with sickle cell anemia related to this research sign an informed consent. A total of 40 Sudanese sickling patient. Sample collected by simple random sampling data collection and measurement, By special designed questionnaire. About three ml was taken in vacuoner tubes in K2 ethylene diamine tetra acetic acid (EDTA) for complete blood counts. Sysmex (hematology analyzer) was used for complete blood counts especially (TWBCs and PLTs) were considered to be measured directly three hydraulic sub systems were used to determine the hemogram; the WBC channel, the red cell, platelet channel and a spritz Hb channel [7]. Differential count: By manual method and blood film was stained with RALL555 stain (modified Geimsa stain) [8]. Data analysis by using SPSS computer software program version 13.

## 3. Results

The age of study population ranged from 8 month to 24 years within the mean age of 7 years as shown in table (1), of the forty study patients 26 were males and 14 females as shown in figure .About 62.5% of Sickler patients have normal differential count while 37.5% have shift to left as shown in table (2). The majority of shift to left sickler (73.3%) have lymphocytosis as shown in table (3), and (26.7%) have neutrophilia as shown in table (4). Association of TWBCs and shift to left as shown in the relation between high total white blood cells and shift to left were statistically significant, with P value (0.0001) as shown in table (5). The majority of patients (60%) are complicated with other diseases like bacterial, viral infection, inflammation and thrombosis as shown in figure (3).The most immature cells present in shift to left patients are band form (93.3%), myelocytes (53.3) and metamyelocytes (26.6%).

**Table 1** showing the distribution of study population by age

Age group	Frequency	Percent
Valid less than 1 years	2	5%
1 _____ 3	11	27.5%
4 _____ 6	10	25%
7 _____ 9	5	12.5%
10 _____ 12	4	10%
13 _____ 15	3	7.5%
16 _____ 18	3	7.5%
19 _____ 21	2	5%
22 _____ 24	0	00%
Total	40	100%

**Table 2** Distribution by shift to left

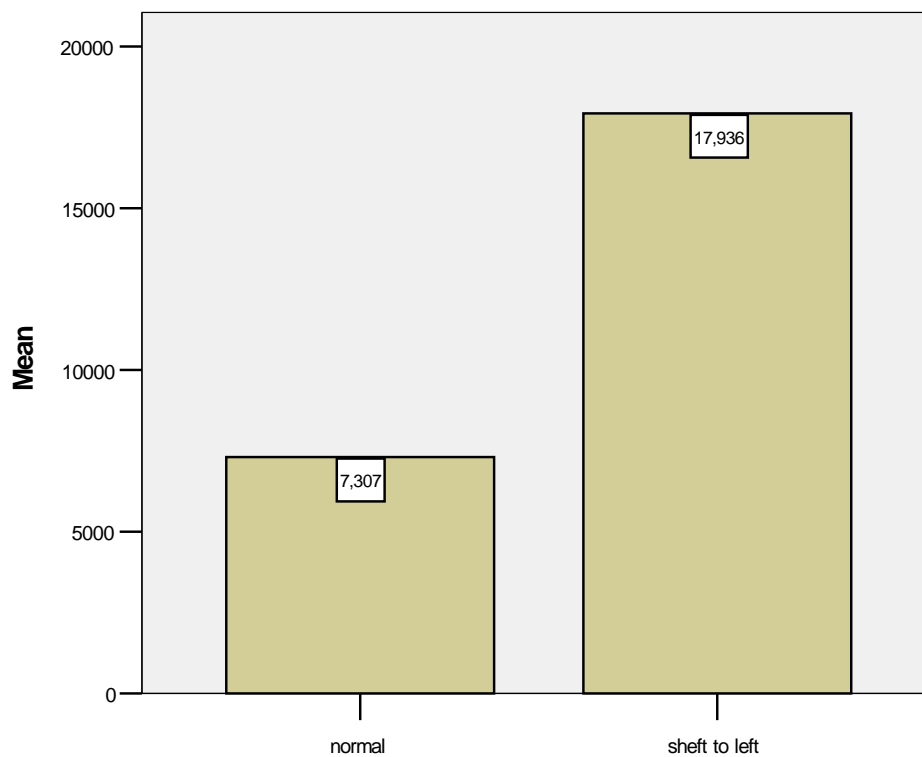
	Frequency	Percent
Valid normal	25	62.5%
Shift to left	15	37.5%
Total	40	100.0%

**Table 3** showing association of shift to left and lymphocytosis

		lymphocytosis			
		Frequency	Percent	Valid Percent	Cumulative Percent
Valid	yes	11	73.3	73.3	73.3
	no	4	26.7	26.7	100.0
Total		15	100.0	100.0	

**Table 4** showing association of shift to left and neutrophilia

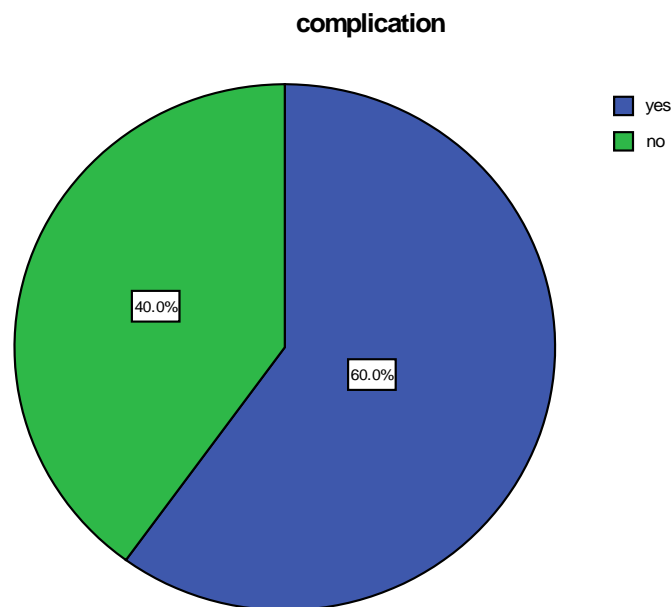
		neutrophilia			
		Frequency	Percent	Valid Percent	Cumulative Percent
Valid	yes	4	26.7	26.7	26.7
	no	11	73.3	73.3	100.0
Total		15	100.0	100.0	



**Figure 2** Association of shift to left and TWBCs

**Table 4** The test of normality showed that the total white blood cells in normal and shift to left was normally distributed.

	Kolmogrove-smimov <sup>a</sup>	Shaprio-wilk
	sig	sig
WBCs		
Shift left	.0001	.0001
Normal	.0001	.0001



**Figure 3** showing that complication may affect on presence of premature WBCs on peripheral blood.

#### 4. Discussion

Sickle cell disease (SCD) is an inherited autosomal recessive disorder with presence of Hb S in blood. This disease affects millions of peoples globally which results in serious complications due to vasoocclusive phenomenon and hemolysis. In this study fifteen results from total of 40 results have been showed increase in total white blood cells. This study agrees with study of Okpala (2004) that has been observed leukocytosis in sickle cell anemic patients which precipitates in pathogenesis of vascular occlusion due to increase in leukocytes adherence to endothelium [9]. Lymphocytosis in Sickler patients indicates immunity against infections but we must continually remember that the lymphocytosis is considered normal in children below 2 years (decrease in neutrophil after period of newborn leaving lymphocytes to increase). Also, the results have been showed neutrophilia; it has been found that when there is an increase in neutrophils, when there is acute infection in the body. In this case the numbers of bands are also increased, and this leads to a decrease in the number of other cells in the blood and this is known as neutrophils left shift. Neutrophilia is a risk factor for a sickle crisis [10–13]. The higher the white cell count, the greater the frequency of acute chest syndrome [14] and stroke [15]. Conversely, reduction of the count in patients with sickle cell disease is beneficial. Some studies have been reported in several study [16]. An elevated baseline leukocyte count is associated with an increase risk of early death, and leukocytosis play a significant role in the initiation of vaso-occlusive events [17]. Several limitations can be highlighted in this study. The study recruited SCD patients who attended a single hospital in Elobied city, Sudan. Also, through a questionnaire, all Sickler patients were found to be using supportive therapy which only improve their life example (folic acid and corticosteroid) these drugs may also make total white blood cells and differential abnormal. Also, there

are many mechanisms in sickle cell disease can change in blood components, blood coagulation during different sickle cell crisis [18].

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## 5. Conclusion

The conclusion of this study there is increase in total white blood cells with shift to left in neutrophil precursor in sickle cell patients with complications ,the most immature cells are band form, myelocytes and metamyelocytes, and there also lymphocytosis and neutrophilia which has been increases in response to infections.

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## Compliance with ethical standards

### *Acknowledgments*

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### *Disclosure of conflict of interest*

All authors have none to declare.

### *Statement of informed consent*

Ethical clearances were obtained from Kordofan University Ethical committee and Informed consent was obtained from all individual participants included in the study

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