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Possible Medical Intervention for Sickle Cell Disease attended to Emergency Room-White Nile State-Sudan

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Abstract

Sickle cell illness included sickle cell disease, sickle cell anemia and other hemoglobinopathies included mutations for sickle cell anemia and others. Here in Sudan, sickle cell illness accumulated in different communities but vast majorities among western population, due to in door marriages, cousins, and so on. Usually lifestyle contributes in health state crisis sickle cell illness patients face, as poverty lead to malnutrition, which has consequences in red cell formation and oxygen tension later, leading to sickling crisis, pain and other complications. This study aimed to assess complete blood count, prothrombin time (PT), INR and D dimer in order to evaluate and provide descent results patients not all the time can conduct due to many reasons, financially the main. 45 sickle disease patients were involved in this study, they were SS and AS Hb types, 20 healthy subjects as control group for comparison of data. CBC conducted via hematology analyzer Mindray-BC3000, PT and INR via Quatron device – Biosystem device and reagents, D dimer through Niycocard device. Data analyzed through statistical package of social science (SPSS0 version 22. Outcome showed low Hb levels among all patients, white blood cell count varies, as well as platelet count. D dimer also fluctuated. Significant differences obtained when comparing data of case group with control group for Hb, WBC and D dimer, the rest did not.

Key word: Sickle cell disease, D dimer, hemoglobinopathies, prothrombin time

1. Introduction

Sickle cell disease (SCD) is caused by the beta-globin gene mutation on chromosome 11, which results from homozygous and compound heterozygote inheritance of a mutation in the β-globin gene. Hemoglobin S (HbS) is the product of a single base-pair point mutation (GAG to GTG) that replaces the hydrophilic amino acid glutamic acid with the hydrophobic amino acid valine in the sixth position of the hemoglobin β-chain [1]. The disease's name originates from sickle cell hemoglobin, which is produced when red blood cells take on a sickle shape. A single gene mutation of the hereditary disease caused an individual to develop sickle cell traits. When a person has two defective hemoglobin genes—either one hemoglobin S gene from one parent and another defective hemoglobin gene, such as beta (β)

thalassemia, hemoglobin C, hemoglobin D, or hemoglobin E, from the second parent—they will develop the disease [2].

Recent global estimates suggest that more than 300 000 affected children are born annually with SCD, about two-thirds of them in Africa [3]. Situated in northeastern Africa, the Republic of South Sudan is regarded as a lower middle-income nation with significant differences between urban and rural areas and consistently poor health outcomes. Malnutrition, poor perinatal outcomes, and persistent infectious diseases linked to poverty place a significant strain on the nation's healthcare system. The health system faces significant problems due to the increasing prevalence of communicable diseases and chronic non-communicable disorders (NCDs). Therefore, avoidable causes like malaria claim the lives of about 78,000 children under the age of five each year. Disease control efforts are undermined by inadequate health care delivery, which is a result of infrastructure inconveniences such inadequate access to laboratories and scarce resources like labor shortages[4].

Sickle cell illness and sickle cell trait differ greatly from one another. While sickle cell trait protects against severe malaria and is generally benign, sickle cell disease is a lifelong, severely incapacitating condition that results in a lower quality of life, high medical resource utilization, increased financial burden, and almost certain early death. Acute and chronic pain, severe anemia, kidney dysfunction, acute chest syndrome, stroke, and other cardiovascular diseases, increased susceptibility to infectious diseases (including malaria), pregnancy complications, and maternal mortality are all consequences of sickle cell disease, which is caused by malformed, sickle-shaped red blood cells that obstruct capillaries and prevent tissue oxygen delivery [5]

Because HbS is less soluble, it can more easily polymerize in hypoxic environments, forming sickle-shaped red blood cells. The primary acute and chronic complications of this disease, such as acute chest syndrome (ACS), splenic sequestration, stroke, pain, and vaso-occlusive crises (VOCs), are caused by this morphological abnormality at the level of the microvasculature, which results in vaso-occlusive phenomena and hemolytic anemia. Repeated artery occlusion at the spleen level results in early hyposplenism and an elevated risk of infection, especially from encapsulated microorganisms [6].

For the purpose of risk stratification of time-dependent priapism-related erectile dysfunction and penile fibrosis, it is crucial to distinguish between major priapism (lasting ≥4 hours) and stuttering priapism (lasting <4-hour episode) [7].

Pain is a common reason why children with sickle cell disease visit the emergency department (ED). Sickle cell illness is characterized by pain, which also dominates children's medical lives. Approximately 70% of hospitalizations for sickle cell disease patients who report to the emergency department are due to uncontrolled pain. The majority of painful episodes in children and adolescents with sickle cell disease do not receive medical attention, despite the perception that patients with sickle cell disease frequently visit the emergency department due to pain. In actuality, 90% of pain episodes are managed at home [8].

2. Method

In this study sickle cell disease patients were involved, as they were in sickle cell disease emergency room a hospital –sections for sickle cell anemia, in White River state, Kostti city. They were 45 set as case group and other 20 healthy individuals set as control group. SCD were diagnosed with hemoglobin electrophoresis, sickling test as conformity for diagnosis, considering the family history and relationship of parents and existence of the disease around. They were assessed for the routine check-up beside the

current complains they were attended the clinic for. Whole blood samples were withdrawing for complete blood count, prothrombin time (PT), INR and D dimer levels as basic tests. Not all tests usually available, so for this study, patients handed results of investigations in order to help in the assessment.

3. Result

This study was conducted as cross sectional case control one, it included 45 patients with sickle cell disease. They have disease in the family descendants, most of them with relative's parents, that what caused existence of the double inheritance of double mutation for both chromosomes. They were from tribes of Western origins. Other 20 subjects were included as control group; they were healthy set for comparisons. Case group appeared with range of age 1 year to 28 years, duration with range 6 months minimum and 48 months as maximum. The control group range ages 2 to 13 years as in table 1. Gender for case group included 44.4% males and 55.6% females, while control group included 50% males and 50% females as in figure 1

Table 1: Descriptive Statistics of Cases and Controls

	N	Minimum	Maximum	Mean	Std. Deviation
Case					
Age	45	1	28	9.0	5.4
Duration/month	45	6	48	11.4	8.5
Control					
Age	20	2	13	7.9	3.4

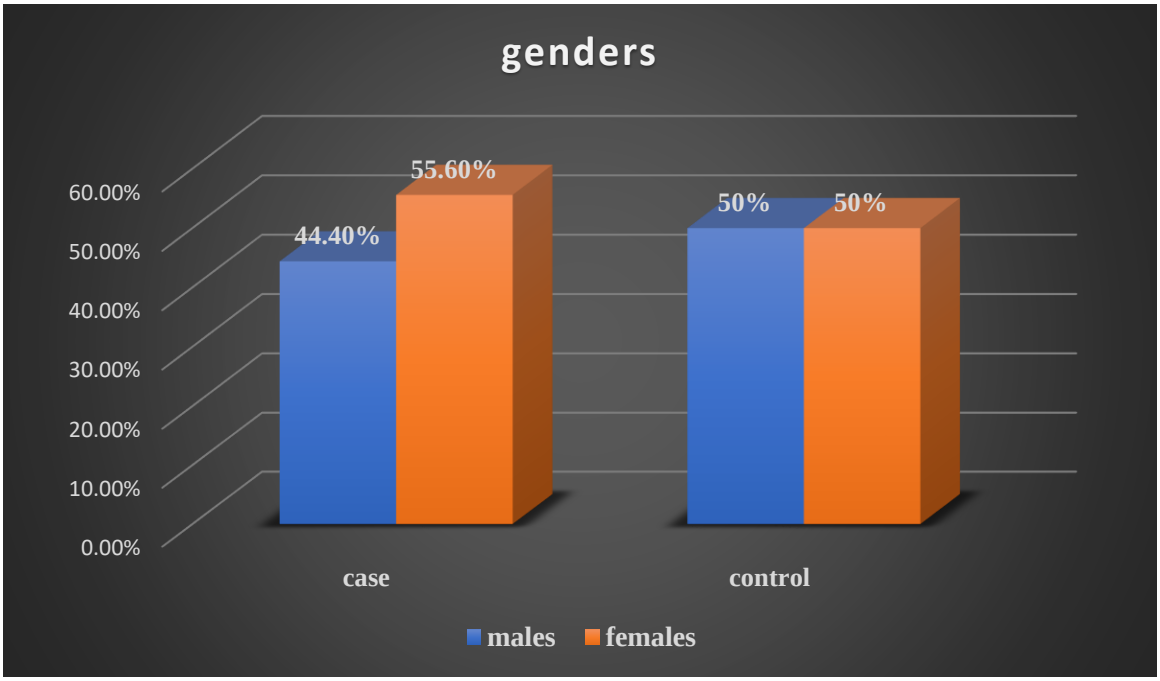

Figure 1: Gender Distribution Among Cases and Controls

SCD patients admitted to the ER according to their diagnosis were with Hb SS (95.6%) and the rest AS were 4.4%. they were presented with crisis with acute pain presented among 46.7%, priapism among 4.4% and the rest with no crisis 48.9% as in table 2.

Table 2: Distribution of Hb Type and Crisis Status

Variable	Category	Frequency	Percent
Hb Type	SS	43	95.6%
	AS	2	4.4%
Crisis	Acute Pain	21	46.7%
	Priapism	2	4.4%
	No Crisis	22	48.9%

Parameters measured were of CBC, PT, INR and D dimer, for both case and control. For case, Hb was ranged form 4.3g/dl to 8.8g/dl, White blood cell (WBC) ranged from 7.8 to 27.3, platelet from 84 to 869, PT 10.5 sec to 17.3 sec, INR 0.8 to 1.4 and D dimer 9.1 to 35.2. readings for control group were normal as I table 3.

Hematological and Coagulation Parameters in Cases and Controls

	N	Minimum	Maximum	Mean	Std. Deviation
Case					
WBC	45	7.8	27.3	14.8	4.4
Hb	45	4.3	8.8	6.4	0.9
Platelet	45	84	869	361.6	170.9
PT	45	10.5	17.3	13.3	1.7
INR	45	.8	1.4	1.1	0.1
D-dimer	45	9.1	35.2	22.3	5.7
Control					
WBC	20	3.8	12.0	7.3	2.7
Hb	20	9.3	20.1	14.6	2.5
Platelet	20	96	957	298.4	189.8
PT	20	11.3	16.2	13.6	1.2
INR	20	.9	1.3	1.1	0.1
D-dimer	20	.2	4.8	0.6	0.2

According to measured parameters, WBC reflection for infection, it was high among 84.4%. level of Hb reflect the anemia occurred, it was low among all of them. While platelet count usually reflect the state of coagulation state, it was increased among 20% and low among 8.9% and the rest were normal as in table 3 and figure 2.

Table 3: Distribution of WBC, Hemoglobin, and Platelet Categories Among Participants

Category	WBC	Hb	Platelet
Low	0 (0.0%)	45 (100.0%)	4 (8.9%)
Normal	7 (15.6%)	0 (0.0%)	32 (71.1%)
High	38 (84.4%)	0 (0.0%)	9 (20.0%)

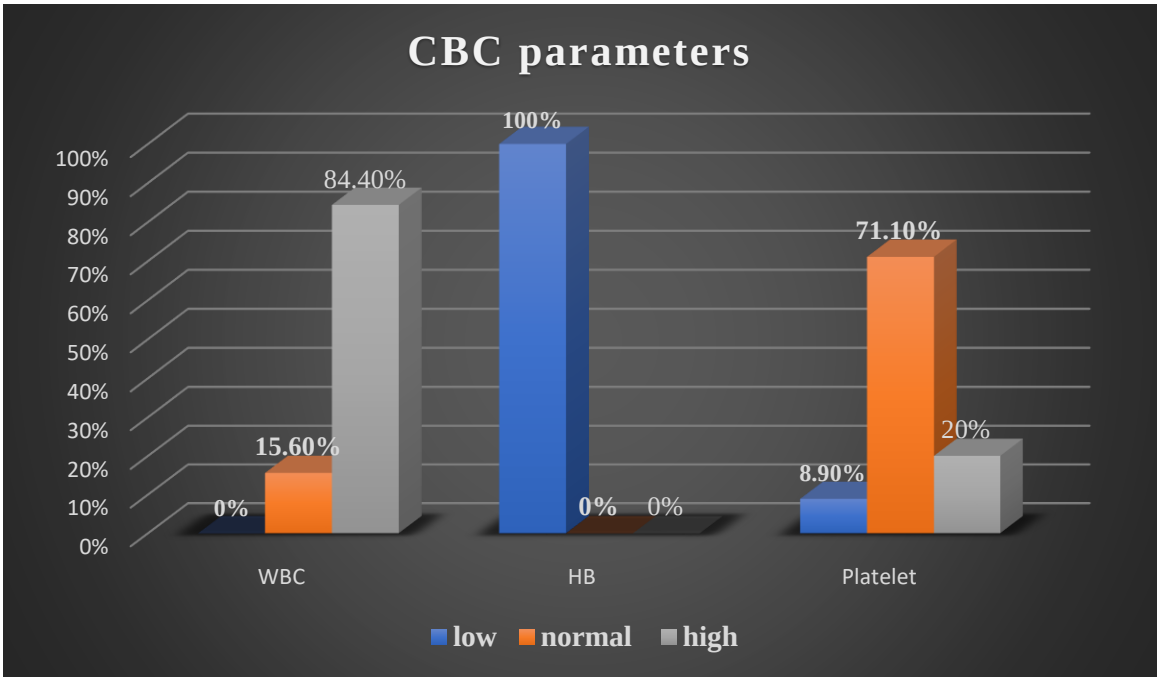

Figure 2: Distribution of WBC, Hb, and Platelet Categories Among Participants

Comparing measured parameters of case with control showed that significant differences for WBC, Hb and D dimer as p value for each was <0.001. the rest platelet and PT and INR showed no changes as p value for each was more than 0.05 as in table 4.

Table 4: Comparison of Hematological and Coagulation Parameters for Cases and Controls

Parameter	Case (n=45)	Control (n=20)	P value
WBC	14.8 ± 4.4	7.3 ± 2.7	<0.001*
Hb	6.4 ± 0.9	14.6 ± 2.5	<0.001*
Platelet	361.6 ± 170.9	298.4 ± 189.8	0.188
PT	13.3 ± 1.7	13.6 ± 1.2	0.535
INR	1.1 ± 0.1	1.1 ± 0.1	0.505
D-dimer	22.3 ± 5.7	0.6 ± 1.0	<0.001*

Significant difference p value <0.05.

Considering gender of Case group, comparing data between males and female's data showed no significant differences obtained as in table 5.

Table 5. Comparison of Hematological and Coagulation Parameters by Gender for SCD

Parameter	Gender		P value
	Male (n=20)	Female (n=25)	
WBC	15.5 ± 3.8	14.1 ± 4.8	0.299
Hb	6.2 ± 0.8	6.5 ± 1.0	0.208
Platelet	362.5 ± 146.8	360.9 ± 191.0	0.977
PT	13.2 ± 1.8	13.5 ± 1.5	0.573
INR	1.1 ± 0.2	1.1 ± 0.1	0.622
D-dimer	22.0 ± 5.4	22.5 ± 6.0	0.754

Considering the type of Hb among SCD patients, comparing measured parameters showed that the only has significant difference was Hb as p value was <0.05, the rest of parameters showed no changes as in table 6.

Table 6: Comparison of Hematological and Coagulation Parameters by Hemoglobin Type in SCD Patients

Parameter	Type of Hb		P value
	SS (n=43)	AS (n=2)	
WBC	14.7 ± 4.4	15.8 ± 4.5	0.749
Hb	6.3 ± 0.9	7.7 ± 0.6	0.032*
Platelet	362.6 ± 174.5	339.5 ± 72.8	0.854
PT	13.3 ± 1.7	14.2 ± 2.1	0.483
INR	1.1 ± 0.1	1.1 ± 0.1	0.734
D-dimer	22.3 ± 5.8	20.6 ± 3.0	0.676

Considering the presence of crisis among SCD patients attended to the ER, comparing measured parameters, no significant differences were obtained as in table 7.

Table 7: Comparison of Hematological and Coagulation Parameters by Crisis Status in Sickle Cell Patients

Parameter	Crisis Status			P value
	Acute Pain (n=21)	Priapism (n=2)	No Crisis (n=22)	
WBC	14.5 ± 3.9	19.7 ± 6.6	14.5 ± 4.7	0.273
Hb	6.3 ± 0.7	6.1 ± 0.6	6.5 ± 1.1	0.579
Platelet	330.2 ± 140.9	353.5 ± 170.4	392.3 ± 197.3	0.502
PT	13.3 ± 1.4	13.8 ± 1.1	13.4 ± 1.9	0.914
INR	1.1 ± 0.1	1.2 ± 0.1	1.1 ± 0.2	0.688

D-dimer	21.2 ± 5.9	25.5 ± 0.0	22.9 ± 5.6	0.446

Pearson's correlation for measured parameters with age of SCD patients showed that, negative correlations with WBC, Hb, platelet, PT, INR and D dimer, but no significant differences obtained as p value for each was >0.05 as in table 8.

Table 8: Correlation Between Age and Hematological/Coagulation Parameters in Sickle Cell Patients

Variable	Pearson Correlation (r)	P value
WBC	-0.247	0.102
Hb	-0.011	0.942
Platelet	-0.113	0.460
PT	-0.055	0.720
INR	-0.117	0.444
D-dimer	-0.069	0.654

Pearson's correlation of duration of the diagnosis of sickle cell disease per months with measured parameters showed that the negative correlation with WBC, platelet, PT without significant differences, positive correlation with Hb with significant difference (p value <0.003) and another positive correlation with D dimer but no significant difference obtained as in table 9.

Table 9: Correlation Between Duration Since Diagnosis (Months) and Hematological, Coagulation Parameters in Sickle Cell Patients

Variable	Pearson Correlation (r)	P value
WBC	-0.228	0.131
Hb	0.431	0.003*
Platelet	-0.292	0.052
PT	-0.126	0.411
INR	-0.132	0.389
D-dimer	0.131	0.390

4. Discussion

In this study SCD presented with anemia, infection and low count of platelets but no signs of vasco-occlusion, crisis presented were manageable. As the resources in the remote areas usually limited, most of inherited disorders have organizations related to the WHO, most of supplements provided through them, beside most if not all SCD were considered poor, so medical and social aids, when provided usually settled in nearest ER. Anemia which presented among all of patients, should reflect the level of liver state, as bilirubin levels due to hemolysis, which usually accompanied SCD patients, but as here in Sudan, the area of residence with low altitude, so no trigger for lower oxygen tension, which leads to

formation of sickle shape, leading to vaso- occlusion, which was not among our patients. But lower Hb concentrations among all patients involved in this study, reflected one of these causes, either bone marrow in aplastic phase. Sickle cell illness is known to cause aplastic and hypoplastic crises. According to recent data, parvovirus infection is the primary cause of the majority of these crises [9]. Another possibility, malnutrition as existence of iron deficiency anemia, low vitamin B12, as in poor food supply causing lazy bone marrow [10]. Also as patients with an active immune/inflammatory response brought on by their present illness have anemia of chronic disease, which results in decreased iron uptake at various sites[11], that can lead to low Hb among SCD.

Splenomegaly, cytopenias (anemia, leukopenia, or thrombocytopenia), and compensatory bone marrow growth are the three components of hypersplenism, the fast and premature loss of blood cells. Hemoglobinopathies and other non-intrinsic splenic disorders cause secondary hypersplenism, such as SCD [12]. Another possible cause of low Hb levels among SCD, out of hemolysis, renal disorders, as it affects production of erythropoietin, therefore decreased production of red blood cell in case kidney disease causing diminished hormone[13].

Usually sickle cell disease managed with the protocol of hydroxyurea, folic acid, iron chelators. comprehensive healthcare management is essential. It includes patient and parent education, appropriate nutrition and hydration, prophylactic antibiotics and folic acid supplementation[14]. To address these issues, a number of therapeutic and preventive strategies are employed. Pneumococcal immunization and hydroxyurea are used to reduce problems, and neonatal screening can help with early diagnosis. A variety of supporting methods to exchange blood transfusions are part of the management of VOCs [15].

Similar studies were conducted around the globe concerning about inherited disorders. A partial agreement obtained with a Nigerian study, in which hematological parameters were assessed for 50 SCD patients with leg ulcer in steady state, and other group without leg ulcer SCD beside 30 healthy subjects as control group. The 80 consenting participants comprised 44 males and 36 females. The average age for SCD cases was 19.58 ± 9.8 years, while controls averaged 27.4 ± 12.3 years. The agreement obtained only in the significant differences were found in hematological parameters between SCD patients and controls, Hb was detected low, WBC was low with significant but in our study it was high than control even with the significant obtained, also the D dimer level of the Nigerian study gave no significant difference unlike our study, and vice versa for PT for our study gave no significant changes [16].

An erection that lasts or happens without regard to sexual desire or stimulation is the hallmark of priapism, a penile erection disease, which presented among 4.4% of patients, reflecting that they were in sickling crisis. While the majority of priapism episodes' end on their own, those that persist more than four hours are regarded as urological emergencies and need to be treated right once. The length of the erection and the degree of tissue ischemia determine the severity of ischemic priapism, which, if left untreated, causes irreparable damage. In order to lower the risk of long-term problems brought on by erectile tissue injury, the main objectives of treatment are to control pain, lower cavernosal pressure, and restore arterial blood flow. Cavernosal aspiration, intracavernosal injections, normal saline irrigation, and surgical shunting are first-line procedures that can be employed to accomplish this goal [17].

5. Conclusion

These study findings reflected the state of sickle cell disease patients. Remote areas usually depend for medical care for hereditary disorders global aid, from WHO mainly and some of the ministry of health

fund usually provided. The severity considered for hemoglobin state, as it was low among all patients participated.

6. Recommendation

Educational programs should be intensified, as the main issue for spreading the hereditary disorders, is the consanguinity marriage. Health state drawing due to poverty most of cases, as population lifestyle reflect the state of homecare they can provide for their patients.

Enhancing for raising lives' standard will provide prospective for general life manners.

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