EVALUATION OF IRON STATUS AMONG SUDANESE PATIENTS WITH SICKLE CELL DISEASE RECEIVING CHRONIC RED BLOOD CELL TRANSFUSION

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ABSTRACT

Sickle cell disease (SCD) is type of haemoglobinopaity and its produced by single base pair change at the 6\textsuperscript{th} codon of the $\beta$ gene followed by replacement of anamino acid Glutamine by valine .Multiple transfusion therapy has been used more frequently to prevent and treat the complications of sickle cell disease. Iron profiles are considered to be of the most important tools in the measurement of the iron balance in steady - state sickle cell disease. Increased Gastrointestinal absorption of iron has been reported in sickle cell disease because of the associated chronic hemolysis, and it is also though that repeated red cell transfusion caused excessive iron level. This study was aimed to evaluate the body iron status in Sudanese patients with sickle cell disease receiving chronic red blood cell transfusion. A total of 50 patients diagnosed with Sickle cell disease attended to Mohammed Elameen Hamid Hospital for children Khartoum, Sudan. Blood samples were collected from each patient, and iron profile was measured using automated chemistry analyzer Mindary (BS 200). Hematological parameters were measured using automated hematology analyzer. Patients’ data was collected using structured interview, questionnaire and from patients’ medical files, and analyzed using statistical package for social sciences (SPSS) version 19. A total of 50 patients diagnosed with Sickle cell disease attended to the Mohammed Elameen Hamid Hospital for children of Khartoum state , Sudan , 24 (48%) of them were males and 26 (52%) were females; their ages ranged from 3-15 years (Mean± SD: 7.5 ±3.3). The result showed that statistically positive  significant association  between serum iron  and number of blood  bags ( P. value : 0.044), and insignificant association with TIBC ( P. value :0.80), serum ferritin(P. value :0.166)  insignificant association with Hemoglobin (P. value :0.726) , Hematocrit (P. value: 0.776), MCV(P. value :0.519), MCH(P. value :0.240), MCHC(P. value :0.208). Serum iron, serum ferritin are significantly decreased and serum total iron binding capacity are significantly increased.

Keywords: Sickle cell disease, Multiple transfusion, Iron.

INTRODUCTION

Sickle cell disease (SCD) refers to a group of disorders caused by autosomal recessive inheritance of a pair of abnormal hemoglobin genes, including the sickle cell gene. It is characterized by chronic hemolytic anemia and acute episodic clinical events called crisis [1].

Vaso-occlusive (painful) crisis is the most common, and other crises are acute hemolytic crisis, sequestration crisis and aplastic crisis [2]. The shortened life span of the sickle red cell coupled with other clinical complication often necessitates frequent blood transfusion in order to maintain a manageable hematocrit [3, 4]. The sickling factors that precipitate the occurrence of sickle cell crisis may be affected by multiple factors such as local tissue hypoxia, dehydration, infections, acidosis, stress and cold.

High levels of fetal hemoglobin are known to ameliorate the severity and incidence of sickle cell crisis and other complication of the disease [5].

This study was aimed at evaluating the iron status of chronic blood transfused sickle Cell Disease patients in Khartoum state, Sudan. Ferritin is a high-molecular-weight protein that contains approximately
20% iron [6].

It occurs normally in almost all tissues of the body but especially in hepatocytes and reticulo endothelial cells, where it serves as an iron reserve. Ferritin is also present in the serum in minute amounts, where it appears to reflect iron stores in normal individuals. Ferritin plays a significant role in the absorption, storage, and release of iron. As the storage form of iron, ferritin remains in the body tissues until it is needed for erythropoiesis. When needed, the iron molecules are released from the apoferritin shell and bind to transferrin, the circulating plasma protein that transports iron to the erythropoietic cells. Transferrin is the plasma iron transport protein, which binds iron strongly at physiological pH [7]. Transferrin is generally only 25% to 30% saturated with iron [8].

In addition to serum ferritin and transferrin, serum iron and total iron binding capacity constitute indices of iron status in human subjects. The aim of the current study was to study all four biochemical markers in an effort to assess the iron status of children with sickle cell anemia. It is expected that the findings would add to available information and assist in patient care.

There is no doubt that both acute and chronic RBC transfusions, both in the acute and chronic setting, can ameliorate many of the complications of sickle cell disease. Some transfusion practices, such as perioperative transfusion, [10] transfusion during pregnancy, [11] and chronic transfusion to prevent stroke in children found to be high-risk from transcranialdoppler (TCD) screening and/or because of previous stroke [12, 13].

**OBJECTIVES**

The purpose of this study was to evaluate the iron status of chronic blood transfused sickle cell disease (SCD) patients referring to Mohammed Elameen Hamid Hospital for children.

**MATERIALS AND METHODS**

**Patient and sample**

This study is a descriptive cross-sectional study, conducted at Mohammed Elameen Hamid Hospital for children, Sudan.

**Study population**

A total of 50 patients diagnosed with Sickle cell disease attending to the Mohammed Elameen Hamid Hospital for children, Khartoum, Sudan, during the period from December 2014 to January 2015 were enrolled to participate in this study.

A 2.5 mL blood sample was collected from each subject into lithium heparin container during the period of steady state. The plasma was separated and stored at −20°C on the day of collection until analysis for estimation of iron profile which was measured using mindray (BS 200, china). Another 2.5 mL of blood was collected in ethylene diamine tetra acetic acid (EDTA) for determination of Hemoglobin (Hb), mean cell volume (MCV), mean cell hemoglobin (MCH), and mean corpuscular hemoglobin concentration (MCHC) by Hematology Analyzer (BC 3000, China) same day of collection.

**Statistical Analysis**

Data of this study was collected by structured interview questionnaire and analyzed using statistical package for social sciences (SPSS) version 19.

**Ethical considerations**

This study was approved by the faculty of medical laboratory sciences Unielain University, and informed consent was obtained from each participant before sample collection.

**RESULTS**

A total of 50 Sudanese patients diagnosed with Sickle cell anemia treated with chronic blood transfusion therapy were enrolled in this study. They were from both gender; their ages ranged between 3-15 years (Mean±SD: 7.5 ±3.3), 24 (48%) of the patients were males and 26 (52%) were females.

The mean of serum iron among studied group was 28.0 and the mean of serum ferritin was 9.0 while the mean of TIBC was 1004. There was statistically significant correlation between serum iron and number of blood bags.

In the SCD patients there was no statistically significant correlation between iron profile and Hb,(P. value :0726) PCV(P. value: 0.776), MCV(P. value :0519), MCH(P. value :0240) , MCHC(P. value :0208) .

The results revealed that (Hb level, Mean±SD: 6.7±1.7 g/dl). PCV , Mean±SD: 20.6±4.19),MCV , Mean±SD: 81.1±9.2 , MCH , Mean±SD: 27.2±3.1,MCHC , Mean±SD: 33.4±2.6, NO of bags transfused , Mean±SD: 11.6±2.3 , serum iron, Mean±SD: 28.1±16.0, serum ferritin ,Mean±SD: 9.0±3.7 , TIBC , Mean±SD: 1004.6±649.9.

The statistical analysis show that low serum ferritin, 24 male (48%) (mean±SD: 8.2±3.0), 26 female( 52%) (mean±SD: 9.7±4.2) ,low serum iron(mean ±SD 9.7±4.2) , (mean ±SD 23.4±8.6) , and high TIBC (mean ±SD 1028.0 ± 566.1), (mean ±SD: 980.2±729.5).

**DISCUSSION**

Many researchers have suggested the effect of iron profile in the patients of sickle cell anemia receiving chronic blood transfusion therapy. This study is a cross sectional study conducted to examine iron profile and hematological parameters with SCD.

Blood transfusion in patients with sickle cell anemia serves two major functions, i.e, increasing the
oxygen-carrying capacity of the blood a [14] and replacing abnormal red cells with normal ones, thereby alleviating symptoms and preventing complications [15,16].

Our result showed that reduced serum iron, serum ferritin was the most common among patients with sickle cell anemia followed by elevated TIBC. This was inconsistent with study done by S Akodu et al which reported that children with sickle cell anemia have higher serum ferritin than controls, implying relatively higher iron content in the reticulo endothelial cells.

**CONCLUSIONS**

All patients may have had multiple transfusions. However, this study did not show a statistically significant correlation between previous history of blood transfusion, serum iron, ferritin concentration and there was statistically significant correlation between TIBC and numbers of blood bags.

**REFERENCES**


