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A HEMATOLOGICAL AND CLINICAL STUDY ON THE SICKLE CELL ANEMIA AMONG CHILDREN

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Abstract:

Background: *Sickle cell disease (SCD) is considered as one of the major types of anemia therefore, we studied clinical and hematological findings to obtain an initial baseline data on sickle cell anemia in children.*

Place and Duration: *In the Pediatric Unit II of Jinnah Hospital Lahore for one-year duration from April 2019 to April 2020.*

Materials and Methods: *Total 75 children were enrolled in the study prospectively using a questionnaire. Clinical data, as well as hematological parameters of full blood count (FBC) for each patient, were carefully obtained.*

Results: *About 80% of the children were initially diagnosed at age less than 5 years. The study showed high frequencies of sickle cell disease among Falatah (13.3% vs 16%) and Jawama (12% vs 13.3%) for fathers and mothers, respectively. Total 80% of children's parents were relatives or from the same tribe. The most frequent clinical presentations were fever (84%), pallor (79%), and bone pain (62%). While common clinical findings were hepatomegaly (44%), joint swelling (35%), and bone swelling (31%). The overall mean hemoglobin concentration was 8.0 ± 2.4 g/dl, packed cell volume was 24.5 ± 7.0 percent, total white blood cells count was 14257 ± 7302 cells/ µL, platelets count was 470821 ± 162245 cells/ µL and reticulocytes count was 12.1 ± 9.8 percent.*

Conclusion: *Epidemiological surveys are needed to determine exactly the magnitude of this problem at least in the region as well as designing a strategic plan to address urgent and long-term interventions in control of sickle cell disease.*

Keywords: *Sickle cell anemia, Children, Hematological, Hospital,*

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INTRODUCTION:

The first description of abnormally elongated red blood cells has been a hundred years since the patient's anemia, and now sickle cell anemia (SCA) has been published combining clinical symptoms called. Sickle cell anemia is an inherited blood disease caused by the replacement of 6 glutamic acids and valine in the globin chain, which will eventually result in an unoxidized sickle hemoglobin to form polymers that destroy red blood cells[1-3]. The remains of the cylinders are located on the outer surface of the hemoglobin and cause intrametric hydrophobia, which reduces the resolution and stability of anaerobic hemoglobin S (HbS). Heterozygote people cause asymptomatic carriers and homozygous sickle cell anemia. The incidence of sickle-blood diseases is higher in sub-Saharan Africa. Although the lack of diagnostic devices means that accurate data are not available, the latest estimates show that more than 230,000 affected children (0.74% of births in sub-Saharan Africa) are born each year, accounting for about 80% of the total number of children in the world. By contrast, the annual estimated birth rate in North America is 2,600 and 1,300 in Europe[4-5]. Sickle blood disease in Africa has been found to be associated with very high infant mortality rates ranging from 50% to 90%, but there is no reliable and up-to-date information. Little is known about sickle disease in Africa. In general, there is a lack of diagnostic devices, there are no routine screening scans, and most patients die undiagnosed in early childhood, despite the fact that financial interventions will survive if provided by a simple package. Sickle disease in Pakistan was first reported in 1926[6-7]. The disease is considered one of the main types of anemia in western Sudan, where the sickle cell gene is particularly common. Research on haemoglobinopathies in Pakistan has shown that hemoglobin "S" was the most common abnormal hemoglobin. Cet has proved to be common among African-speaking groups from Asia, including Arab and non-Arab nomadic groups migrating to Sudan during various historic crossings[8-9]. Infants with HBS rarely have complications in the first months of life, while hbf levels remain high. The disease develops clinically from the third month, where the first presentation is usually caused by painful swelling of the hand or the back of the foot of the syndrome "foot", usually a symmetrical, small myocardial infarction. Patients suffer from chronic ill health interspersed with acute anemic disease, heart attacks and infectious crises[10-11]. The purpose of this study is to obtain the first clinical and hematological reference data on sickle cell anemia in children.

MATERIALS AND METHODS:

The study was conducted in the Pediatric Unit II of Jinnah Hospital Lahore for one-year duration from April 2019 to April 2020. Participants were children with sickle cell anemia during their work period. The reference point for inclusion has been diagnosed by all older children (0-15) years old and their doctor as a patient with sickle cell disease. A total of 75 sickle-blood children were enrolled in the study. The working protocol was approved by the Ethical committee. Informed consent was given from each caregiver before taking the sample, and the doctor completed a survey for each participant before the tests were conducted. Anticoagulated venous EDTA blood inflammation was performed and full cadaver morph tests were conducted and results were recorded. All data was entered into the computer and analyzed using SPSS.

RESULTS:

There were 75 children among the study group, 34 (45%) were girls and 41 (55%) were boys. Their age range was 3 months up to 15 years, 1-5 years age group constituted the majority, 39 (52%). Mean weight was 15.8 ± 11.0 kg, while heights mean was 93.3 ± 8.0 cm. Total 80% of the children were initially diagnosed at age less than 5 years. The majority of the children (80%) had completed the routine vaccination according to their age. The study demonstrated that 57% of them were in long-term use of folic acid.

Figure 1 The major presenting symptoms of admitted children with SCD

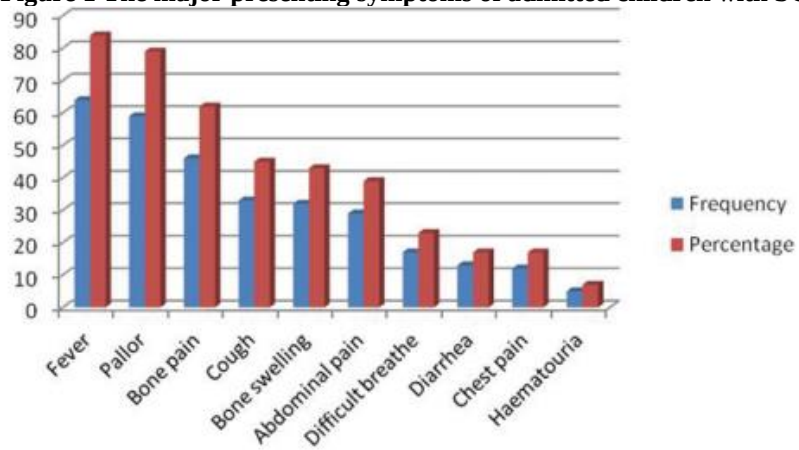
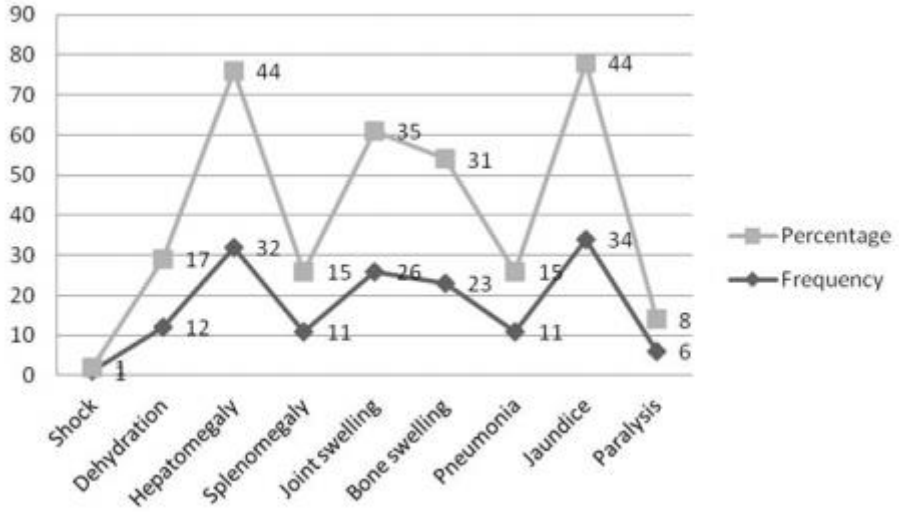


Figure 2 The major signs seen in admitted children with SCD,



Due to unavailability of Hb electrophoresis test during the study period, laboratory diagnosis of all cases was based on clinical grounds and sickling test, 84% demonstrated positive sickling test. The overall mean hemoglobin concentration was 8.0 ± 2.4 g/dl, packed cell volume 24.5 ± 7.0 percent, total white blood cells count 14257 ± 7302 cells/µL, platelets count 470821 ± 162245 cells/µL and reticulocytes count 12.1 ± 9.8 percent. When hematological parameters were categorized, results among patients under study were: 39% had hemoglobin concentration 6-8 g/dl, 32% had packed cell volume, 20-25% had total white blood cells count (WBC15000 cells/µL. Total 59% of patients showed elevated platelets count, above 600,000. Reticulocytes count was high (above 5%) in 65% of patients.

DISCUSSION AND CONCLUSION:

It is suggested that HBS is more common in the populations of Pakistan. Although many studies have been conducted in the Kordofan region, most of the data is unpublished. In this article, we try to provide basic data on clinical, hematological and ethical problems associated with sickle disease. Similar results of a report in a study conducted in the same region. Osman, et al., reported the highest frequency in the operation of the Masaleet tribe in algadaref province[12-13]. Ahmed, et al., considered the West Kordofan Misseria tribe to be the most common tribe with HbS, the incidence of sickle disease (SCD) was up to 30%. The same findings are Nimir et al. Studies have shown that most mothers are relatives or of the same tribe, this high frequency of blood marriage has a deep root in their traditions. Blood marriages are also associated with numerous abnormal HB subtypes. As a result, the risk of persistent HB anomalies for the next generation also increased. Designing control measures in these communities is a challenge. 84% of participants were diagnosed with sickle cells, but only 37% had sickle disease in the past, depending on the family[14]. This is because many families deny this disorder so that they are not stigmatized. Hbs gene approval sometimes leads to internal conflicts and social problems. When assessing the presentation of symptoms, fever was the main feature (84%), followed by pallor, bone pain. This is consistent with bayoumi, and others reported who mentioned "Usually Sudan patients often have severe anemia, hand and foot syndrome, fever, jaundice and vaso-occlusion seizures." Interestingly, in some cases (10%) hand and foot syndrome, but negative studies of the disease were conducted and hematological parameters differed from patients with sickle cells. More research was needed to accurately diagnose these cases. Joint clinical results were found in the paleness test (79%), hepatomegaly and jaundice (44%), joint swelling (35%), bone swelling (31%), and herring enlargement at just 15%. Osman, et al. Paleness (97%), jaundice (32%), palpable liver (60%), palpable spleen (6%), heart symptoms (27%), hand and foot syndrome (16%). This study showed that 88% of them were bayoumi et al. In their study, all patients had mild to moderate heart expansion; Forty-two% were moderately enlarged spleen, but only 10% had an enlarged liver. The total mean hemoglobin and PVC were 8.0 g/dl and 24.5% respectively[15]. The average number of retinal inflammation was 12.1%. Leukocytosis is definitely associated with poor prognosis, while reducing the number of neutrophils is associated with a good prognosis. Most patients had more than 600,000 relatively high platelets. It's the same as Osman discovered. The negative effect of feedback on erythropoietin production in patients as a result of anemia may be responsible for platelets. Erythropoietin has a structural homology with thrombopoietin, but the second is much larger than the first, but about half of thrombopoietin has a similarity or identity with erythropoietin in the N-

terminal region. For this reason, platelets are well known to be associated with chronic disease anemia and various types of anemia. The decrease or absence of spleen platelet growth as a result of inaction in sickle cell disease contributes significantly to the increase in the average number of platelets in sickle cell disease.

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