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# Sickle Cell Syndrome in Children in Basrah

**Imad A. Ali, Mea'ad K. Hassan**

**Department of Pediatrics, College of Medicine Basrah University, Basrah, Iraq**



## Abstract

*A total of 206 Patients with sickle cell syndrome admitted to Basrah General Hospital from February 1992 till the end to January 1993 were included, their ages ranged from 6 months to 15 years. They were divided into three groups according to their hemoglobin electrophoresis type: Hb – SF group (111 patients), Hb- ss group (89 patients) and Hb- As group (6 patients). Most of admitted cases were in the summer time (August), the second peak of admission was in January. The mean hemoglobin level was 7.5 g/dl in the first group with Hb- SF, 8.27 g/dl in patients with Hb – SS and 11.6 gm/ dl in patients with Hb – AS. Painful crises were the commonest type of crises (41%), followed by hyperhemolytic crises (8%), and aplastic crises (7%). Infections were detected in 21% of patients, pneumonia was the commonest from. Three patients died during conducting the study 91.5%), main causes of death were infections and vaso-occlusive crises.*

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## Introduction

Sickle cell syndrome is one of the important health problems among children in Basrah. The first reported case in Iraq was by Bakir and Al-Qaysi in 1964 [1], it is interesting to mention that the patient in that report was from Basrah.

The expression of the disease at the clinical level is extremely unpredictable and variable between individuals of one race and this variation becomes more evident among different races.

Sickle cell anemia becomes evident clinically in early childhood, and is accompanied by signification morbidity. The clinical course is characterized by periods of remissions and exacerbations (crises), the commonest type is vaso-occlusive (painful) crises. Patients with sickle cell syndrome are more liable for bacterial infections due to several abnormalities of the body's defense mechanism. Bacterial infections are the leading cause of death in sickle cell disease in children, these include pneumonia, osteomyelitis and general sepsis [2].

The aim of the study was to highlight the effect of sickle cell syndrome on children of Basrah.

## Patients and Methods

206 patients with sickle cell syndrome, their ago ranged from 6 months to 15 years, admitted to Basrah General Hospital from February 1992 till the end of January 1993 full information about their history and clinical presentation were obtained. For every patient, complete clinical examination and follow up for the progress of the

disease and complications, in addition important investigations like hemoglobin electrophoreses (using cellulose-acetate with sodium barbital as a buffer solution pH 8.2 – 8.6), radiological, biochemical and microbiological investigations were done.

## Results

Total number of cases included in the study was 206 (115 male and 91 female), male to female ratio was (1.26). These patients were divided into three groups according to the type of hemoglobin on electrophoreses as shown in table (1). The mean hemoglobin level for patients with Hb-SS was 8.27 g/dl, for patients with Hb-SF was 7.5 dm/dl, and for patients with Hb-AS was 11.6 g/dl as shown in table (2). The age of patients was ranging from six months to fifteen years, they were grouped into four groups according to their age as shown in table (3). Most of cases were admitted during the summer time (especially August) and other peak of admission was during January as shown in figure (1).

***Table 1:*** Distribution of patients according to their hemoglobin type.

| Hb type | No. | % |
|---|---|---|
| SF | 111 | 53.9 |
| SS | 89 | 43.2 |
| AS | 6 | 2.9 |

***Table 2:*** Mean hemoglobin level (g/dl) according to age and hemoglobin type.

| Age (years) | Mean Hb level (g/dl) | | |
|---|---|---|---|
| | Hb-SF | Hb-SS | Hb-AS |
| <1 | 7 | 8.2 | |
| 1-5 | 7.4 | 7.8 | 11.2 |
| 6-10 | 7.6 | 8.5 | 12.1 |
| 11-15 | 8 | 8.6 | |
| ***Mean*** | ***7.5*** | ***8.26*** | |

***Table 3:*** Distribution of patients according to their age and hemoglobin type.

| Age (years) | Hb type | | | | | | *Total* | |
|---|---|---|---|---|---|---|---|---|
| | SF | | SS | | AS | | | |
| | No. | % | No. | % | No. | % | *No.* | *%* |
| <1 | 10 | 9 | 7 | 7.8 | - | - | ***17*** | ***8.2*** |
| 1-5 | 34 | 30.6 | 25 | 28.9 | 5 | 83.3 | ***64*** | ***31*** |
| 6-10 | 43 | 38.7 | 37 | 41.5 | 1 | 16.7 | ***81*** | ***39.3*** |
| 11-15 | 24 | 21.6 | 26 | 22.5 | - | - | ***44*** | ***21.4*** |
| ***Total*** | ***111*** | ***100*** | ***89*** | ***100*** | ***6*** | ***100*** | | |

Age specific blood transfusion rate for patients with Hb-SF older than six years was significantly higher than for patients with Hb- SS of the same age group as shown in table (4).

***Table 4:*** Age specific blood transfusion rate between patients with Hb-SF and Hb-SS.

| Age (years) | Percentage of patients who received blood | | p value |
|---|---|---|---|
| | Hb-SF | Hb-SS | |
| <1 | 60 | 28.5 | >0.05 |
| 1-5 | 52.9 | 28 | >0.05 |
| 6-10 | 65.1 | 32.4 | <0.01 |
| 11-15 | 75 | 3 | <0.01 |
| ***Total*** | ***63.1*** | ***30.3*** | ***<0.01*** |

Three forms of sickle cell crisis have been recognized in the study, these include:-

1- Painful crises ⇒ 82 patients 41%.
2- Hyperhemolytic crises ⇒ 16 patients 8%.
3- Aplastic crises ⇒ 14 patients 7%.

Patients with Hb-SS had a significantly higher incidence of painful and a plastic crises than patients with Hb-SF as shown in table (5). Sequestration crises have not been recorded in the study. The most common form of painful crises was generalized pain all over the body in 39 patients (47.5%), other forms of painful crises are shown in table (6). Infections were detected in (21%) of patients with sickle cell syndrome, pneumonia was commonest type of infections in the study, other forms of infections are shown in table (7), statistically there was no significant difference in the distribution of infections among patients with Hb-SF and patients with Hb-SS, also there was no significant difference in the distribution of infections in both types of hemoglobin among different age groups as shown in table (8). There patients died during conducting the study, one patient with acute gastroenteritis and severe dehydration, the second with septicemia, and the third patient died within two days after developing convulsions and left-sided hemiplegia.

***Table 5:*** Types of sickle cell crises in relation to the type of hemoglobin.

| Type of crisis | Hb type | | Total (no. 200) No. (%) | p value |
|---|---|---|---|---|
| | Hb-SF (no. 111) No. (%) | Hb-SS (no. 89) No. (%) | | |
| Painful | 34 (30.6) | 48 (53.9) | 82 (41) | <0.01 |
| Hyperhemolytic | 9 (8) | 7 (7.8) | 16 (8) | >0.05 |
| Aplastic | 6 (5.4) | 8 (8.9) | 14 (7) | <0.01 |
| ***Total*** | ***49 (44)*** | ***63 (70.8)*** | ***112 (56)*** | ***<0.01*** |

***Table 6:*** Distribution of painful crises.

| Type of crisis | No. | % |
|---|---|---|
| Generalized pain all over the body. | 39 | 47.5 |
| Backache | 18 | 21.9 |
| Long bone and joint pain | 13 | 15.9 |
| Abdominal pain | 11 | 13.5 |
| Hand-foot syndrome | 1 | 1.2 |

***Table 7:*** Distribution of different types of infections in relation hemoglobin type.

| Infection | Hb type | | Total (no.200) |
|---|---|---|---|
| | Hb-SF (no. 111) | Hb-SS (no. 89) | |
| Pneumonia | 6 | 9 | 15 |
| Typhoid fever | 9 | 2 | 11 |
| UTI | 2 | 4 | 6 |
| Meningitis | - | 2 | 2 |
| Hepatitis A | 2 | - | 2 |
| Hepatitis B | 1 | - | 1 |
| Osteomyelitis | - | 1 | 1 |
| GIT infection | 2 | 1 | 3 |
| Mumps | 1 | - | 1 |
| *Total* | *23 (20.7%)* | *19 (21.3%)* | *42 (21%)* |

p value >0.05

***Table 8:*** Distribution of infections among different age groups of patients in relation to hemoglobin type.

| Age (years) | Hb-SF | Hb-SS | Total | p value |
|---|---|---|---|---|
| <1 (no. 17) | 2 | 1 | 3 | >0.05 |
| 1-5 (no. 59) | 10 | 9 | 19 | >0.05 |
| 6-10 (no. 80) | 9 | 7 | 16 | >0.05 |
| 11-15 (no. 44) | 2 | 2 | 4 | >0.05 |
| ***Total*** | 23 | 19 | 42 | |

## *Discussion*

This study was a prospective study done over one years on sickler children who were admitted for the following reasons:-

a-Correction of anemia by blood transfusion.

b-Management of sickle cell crises.

c-Management of infections and other complications of the disease.

The two previous studies done in Basrah at 1990 and 1981 were conducted on adult patients mainly and they did not determine the types of complications occurring with this syndrome [3,4].

The study showed that more males than females were admitted to the hospital (1.26/1), we were unable to explain this but studies from neighboring countries [5] and previous studies in Basrah [3,4] showed similar results. More than half of the patients in the study had been shown to have Hb-SF (53.9%), while (43.2%) of patients had Hb-SS and only (2.9%) had Hb-AS, this may be attributed to that these children with Hb-SF have a lower hemoglobin level as shown in table (2) and there need frequent admission to the hospital for blood transfusion. 39.3% of patients included in the study belong to the age group 6 to 10 years, 31% belong to the age group 1 to 5 years, (21.4%) belong to the age group 11 to 15 years and (8.2%) were infants. This may reflect the fact that the manifestations and complications of sickle cell syndrome are predominant in school age group, while in Africa and Jamaica, most of cases have manifestations and complications of the disease at infancy and early childhood [6,7]. In Jamaica 18% of patients with Hb-SS presented before their first birthday [8], while in our study only (7.8%) of patients with Hb-SS presented at infancy. Maximum number of admissions was during the summer time particularly in August and there was another peek during January. These seasonal changes were related mainly to three factors:-

a-Extremes of temperature have been shown to be one of the precipitating factors of vaso-occlusive crises.

b-Dehydration which is common during summer, may be an important precipitating factor for vaso-occlusive crises.

c-Infections:- during summer, there is increase in the incidence of gastroenteritis and typhoid fever, while during winter, there is increase in respiratory tract infections.

In the study, the mean hemoglobin level for patients with Hb-SS was 8.2 g/dl and for patients with Hb-SF was 7.5 g/dl. In African study [8], mean hemoglobin level for patients with Hb-SS was 8 g/dl. And for patients with Hb-SF was 10.3 g/dl, this can be explained by the fact that Hb S/B thalassemia is a heterogeneous group of disorders whose clinical features depend on the severity of the B-thalassemia component, in general they range from the more severe B° form to

the very mild $B^+$ variants [2]. Percent distribution of patients who had received blood during their admission showed that children with Hb-SF had received blood significantly more than children with Hb-SS disease, so these indicate that anemia is more severe in children with Hb-SF.

Three main crises were recognized in the study:-

a- Painful crises (vaso-occlusive) 41%
b- Hyperhemolytic crises 8%.
c- Aplastic crises 7%.

Painful crises and a plastic crises were significantly higher in children with Hb-SS than children with Hb-SF. Painful crises occur in about 72.6% of African patients with Hb-SS [8]. Only one case with hand – foot syndrome (1.2%) was reported in the study, while the incidence of hand-foot syndrome in African sicklers was 28% in patients with Hb-SS [8]. Hyperhemolytic crises which were seen in our patients were due to the co-existence of glucose-6-phosphate dehydrogenase deficiency with sickle cell syndrome. No case with sequestration crises was recorded in the study, this is going with studies in Eastern province of Saudi Arabia where there is very low incidence of splenic sequestration [9], 21% of patients with sickle cell syndrome in the study were shown to have infection, there was no significant difference in the distribution of infections between patients with Hb-SF and Hb-SS.

Pneumonia was the most common form of infection recorded in they study (7.5%) as in other parts of the world [10].

Mortality rate in the study was 1.5% their age were between 3 to 4 years. In Jamaica, the highest mortality rate was during the first five years (10% during the first year, 5% during the second year and 3% during the third year) [11]. The main cause of death was infection.

From this study we conclude that sickle cell syndrome generally runs a milder course in children in Basrah as compared to African, and Jamaican children taking in consideration the following features:-

1- The frequency of crises is comparatively low and dangerous crises like sequestration crises were not recorded.
2- Low mortality rate.

However, sickle cell anemia is still hazardous to our children and form one of the major health problems.

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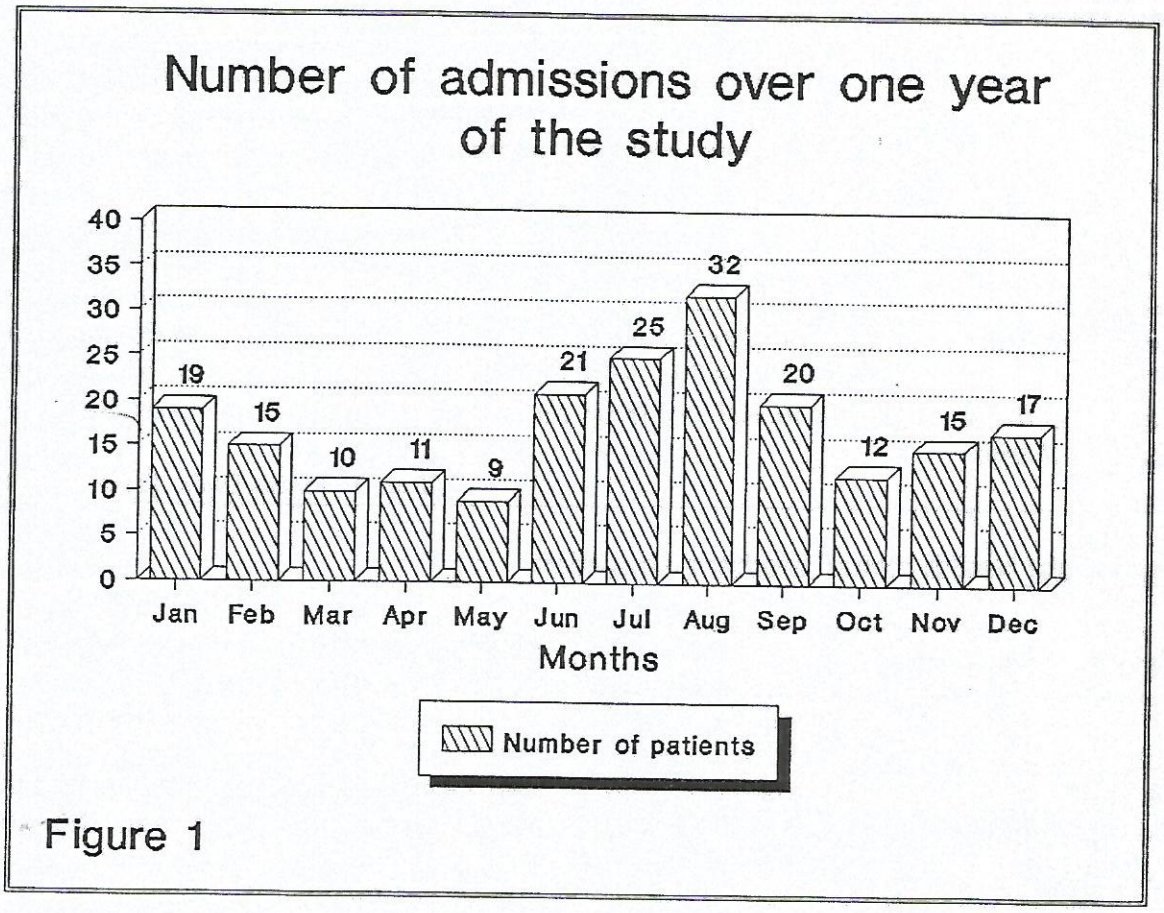


Figure 1