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Original Research Article

TO STUDY THE CLINICAL PROFILE OF PATIENTS WITH SICKLE CELL DISEASE BETWEEN 01 TO 18 YEARS OF AGE AT A TERTIARY CARE CENTER IN SAIMS INDORE (MP) CENTRAL INDIA

Dr. Swati Patel Jain[1] (Assistant Professor), Dr. Priyanka Singh[2] (Junior Resident 3rd Year), Dr. K.K Arora[3] (Professor), Dr. D. Barua[4] (Professor) & Dr. Gunjan Kela[5] (HOD and Professor)

Department of Paediatrics, Shi Aurobindo Medical College and PGI Indore M.P India[1,2,3,4&5]
Corresponding Author: Dr. Priyanka Singh

ABSTRACT

The aim of the study is to study the clinical profile of patients with sickle cell disease between 01 to 18 years of age at a tertiary care center. A detailed history was elicited from parents and patients and clinical examination was done. The risk factors associated, clinical and investigation details, treatment received was noted. Patients diagnosed as sickle cell disease through appropriate laboratory techniques were registered for detail clinical assessment and follow up. The most common clinical features in sickle cell diseases were considered fever (82.9%), followed by pain, pain were classified as abdominal pain (29.2%), chest pain (53.65%), Pain over limb (hand & foot syndrome) (70.7%) and average duration of pain was three days. (17.07%) were founding limping & dyspnea (24.39%) & 65.29% were found to have jaundice during the sickle cell anemia episode. Most of the sickle cell anemia were frequently associated with anemia.

1. INTRODUCTION

Sickle cell disease is an inherited blood condition that is most common among people of African, Arabian and Indian origin that is caused by a single nucleotide mutation that substitutes glutamic acid for valine in the sixth position of the β-globin gene. During hypoxic conditions, the red blood cell becomes sickled, and the resulting change in structure restricts circulation causing obstruction of the blood flow within the capillaries and early destruction of the cell. The awareness about sickle cell disease is highly restricted and is generally diagnosed after the incidence of one of its many complications. Many times death occurs before the disease can be diagnosed, and parents do not have a notion about the life-threatening crises that their child may suffer from. Hence, it has become immensely important to educate the masses to further reduce the mortality and morbidity associated with sickle cell disease. A definitive cure is not currently available for patients with sickle cell anemia (SCA). Existing therapies are only focused on symptom management and do not alter the natural history of the disease. These therapies are comprised of hydration, prevention of

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infections, pain management, proper nutrition, and precautions against adverse weather conditions. Tab Hydroxyurea is an FDA-approved drug used in the management of sickle cell disease and has been found to be an effective therapy, as it reduces the number of hospitalisations, decreases mortality, and improves the quality of life.

Bone marrow transplantation is curative for SCA, but there is a lack of donor availability, as well as long term adverse effects of a bone marrow transplant. In India, the disease is largely undocumented. Thus, there is an urgent need to throw light upon the features of disease so that locally appropriate models of care may be evolved.

2. MATERIAL AND METHOD:

STUDY DESIGN

A cross-sectional study was conducted in the Paediatrics wards of a tertiary care hospital Sri Aurbindo Institute of Medical Science from September 2022 till date. A detailed history was elicited from parents and patients and clinical examination was done. The risk factors associated, clinical and investigation details, treatment received was noted. Patients diagnosed as sickle cell disease through appropriate laboratory techniques were registered for detail clinical assessment and follow up. The clinical history, various investigations are recorded in structured proforma.

The patients were examined clinically as per structured proforma. All patients were given folic acid to be taken daily. Patients and/or their parents were advised to avoid disease precipitating factors like exposure to extreme climate, excessive dehydration, excessive physical stress etc. Patients were also told to seek appropriate medical intervention quickly upon any minor ailment. Patients were advised to take enough water/fluids. Patients were given folic acid, B-complex and anti-pyretic/anti-inflammatory tablets. They were given symptomatic treatment as outdoor patients and indoor patients. The severity of the disease was assessed by converting the clinical observations in to the numerical score.

3. RESULT:

Total 41 patient (Table no1) included in this study, Majority belongs to lower class (87.8%). Majority of patients were male 23 (56.09%) and female patients were 18 (43.9%) (Table no 2).

Age Distribution: (Table no 3) In this study children's less than 18 years were considered, and their manifestation were mostly in age group less than 15 years of age. There is seen less proportion in growing age. Thus sickle cell disease is a medical condition majority it is found in early childhood.

Clinical features: (Table no 4). The most common clinical features in sickle cell diseases were considered fever (82.9%), followed by pain, pain were classified as abdominal pain (29.2%), chest pain (53.65%), Pain over limb (hand & foot syndrome) (70.7%) and average duration of pain was three days. (17.07%) were founding limping & dyspnea (24.39%) & 65.29% were found to have jaundice during the sickle cell anemia episode. Most of the sickle cell anemia were frequently associated with anemia. Majority of case presented with fever

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(without any specific cause) with severe anemia. Such episodes in a sickle cell prone area arouse the suspicion of sickle cell disease. The cause of aggravation of anemia in this situation may be hyperhemolysis. Recurrent severe anemia without evidence of preceding hyper hemolysis was also seen in many children the cause of which is not known. But acute severe anemia with rapidly enlarging liver & spleen. Pain abdomen was a frequent phenomenon in children was due to sequestration. Therefore blood transfusion was given to the children to reducing the chance of mortality in children. Other clinical features like epistaxis, hematuria, stroke, gall bladder, and priapism were not likely found in this studies. In this patients presented with SCD among them were (80.40%) individual had a past history of blood transfusion and average age of first episode of SCD was four year or less majority of them need hospitalisation. Among the patient was 68.29 % had a history of jaundice with an average of three episode & hepatosplenomegaly. Total hemoglobin (fig no 1) was between 06 to 11 gm/dl in most of children 70.73%, less than 6 gm/dl were 17.07% & more than 11 gm/dl were 12.19% with this level of hemoglobin patient thrived & grew well. Most of the patient were symptomatic & treated symptomatic. Patient had been treated with T. Hydroxyurea, Tab folic acid & blood transfusion. In most of the patient fetal hemoglobin HbF was 5-33% (92.68%) and HbF 5% less was (07.3%).Fig No2.

About 78.04% of patients required hospitalisation some time or the other. Hospitalisation was required for treatment for attacks of pain, severe anemia or requiring blood transfusions.

Table No 1: (N 41)

S no.	Income group	No. of patients	Percentage (%)
1	Lower class	36	87.8
2	Upper lower class	4	9.7
3	Lower middle class	1	2.4

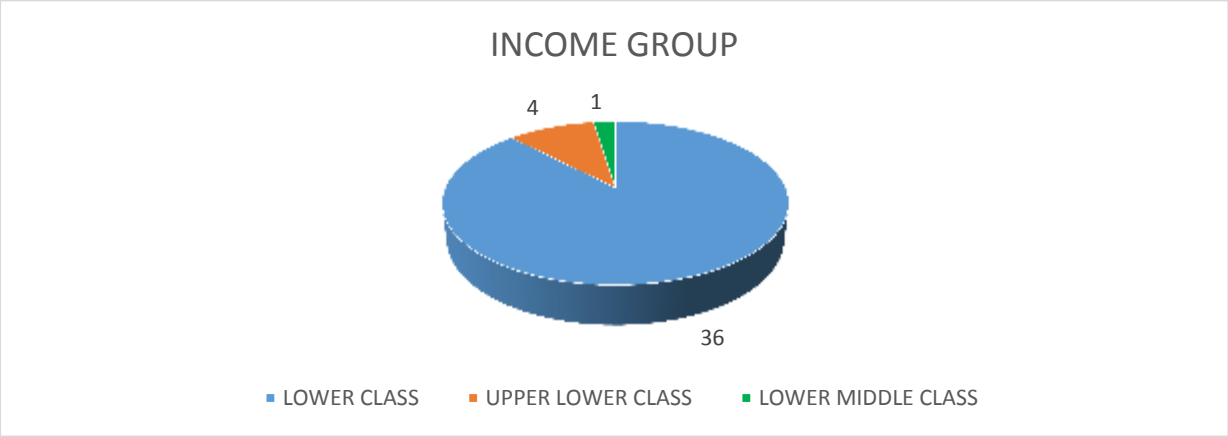

Table No 2:

S no.	Sex	No. of patients	Percentage (%)
1	Male	23	56.09
2	Female	18	43.09

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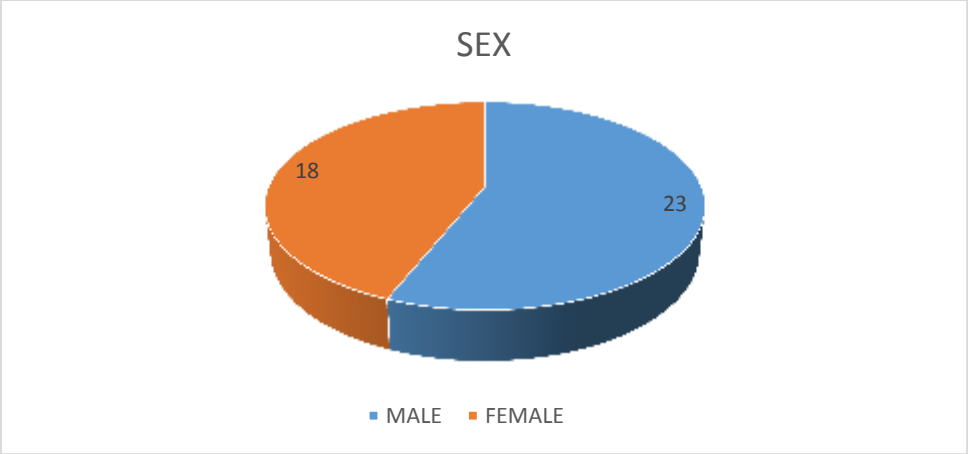

Table No 3:

S no.	Age- group	No. of patients	Percentage (%)
1	0-5 yrs	11	26.82
2	5-10 yrs	15	36.58
3	10-15 yrs	13	31.70
4	15 -18 yrs	2	04.87

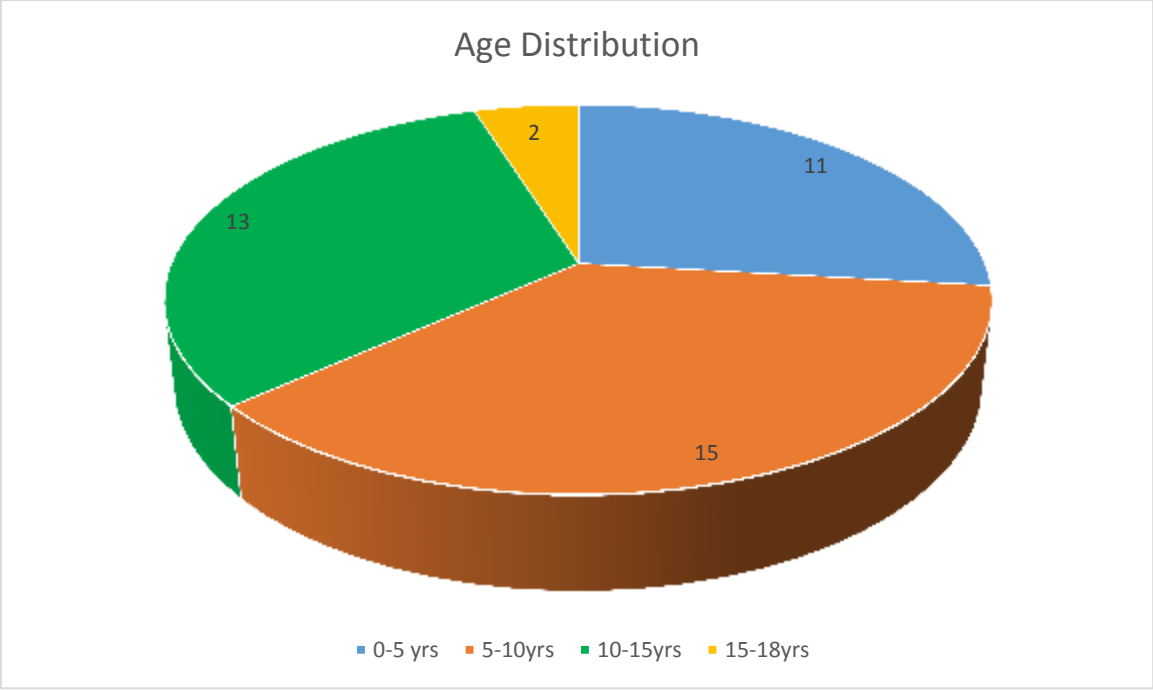

Table No. 4:

S no	Clinical Features	Percentage(%)
1	Fever	82.9
2	Abdominal pain	29.2
3	Chest Pain	53.65
4	Pain over limb	70.7
5	Duration of pain	(average 3 days)

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6	Frequent infections	36.58
7	Limping	17.07
8	Dyspnea	24.39
9	Jaundice	68.29

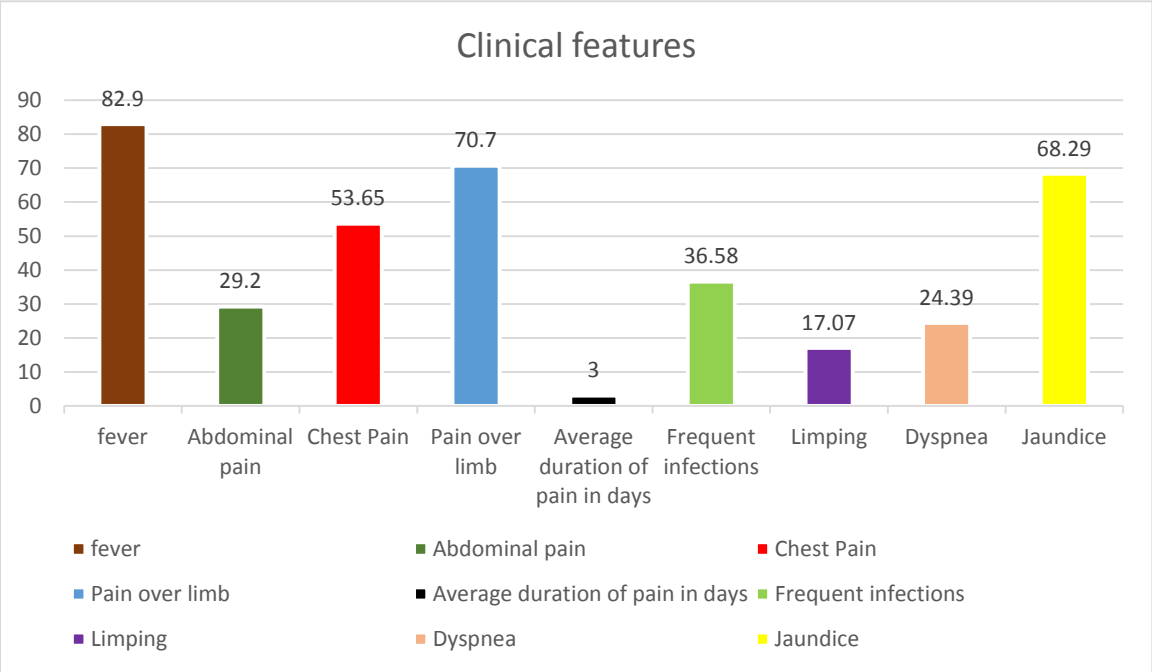

Fig no. 1: (Hemoglobin Hb)

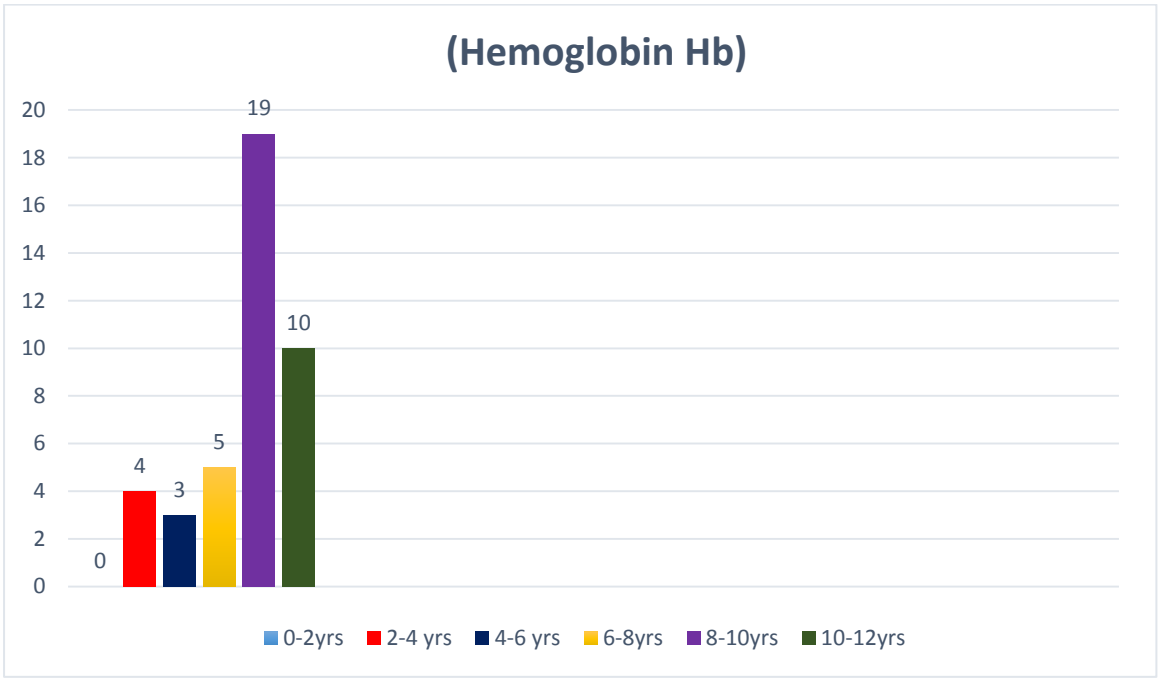

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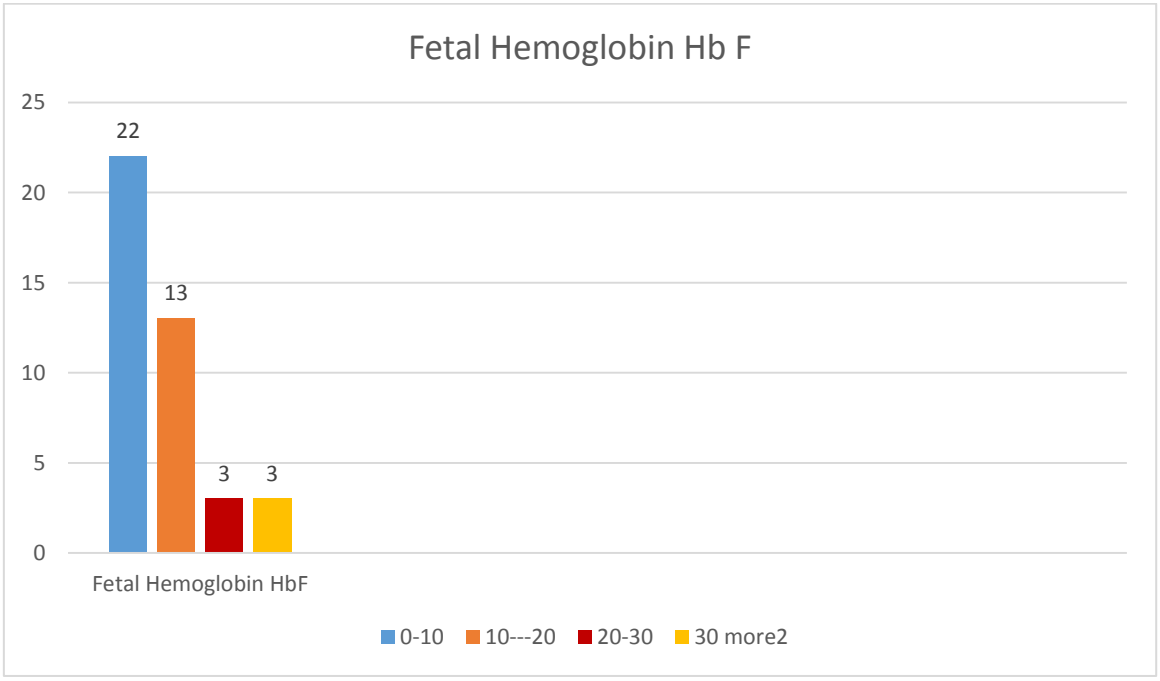
Fig No 2: (Fetal Hemoglobin HbF)



Ethical consideration:

Ethical clearance for conducting the study was obtained from the Institutional Ethics Committee. Information obtained during the study is confidential. Written informed consent in the local language was obtained from the parents or guardians of each enrolled patient who were willing to get enrolled in the study after explaining to them the nature of the study.

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