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STUDY OF HEMATOLOGICAL PARAMETERS IN CHILDREN WITH SICKLE CELL DISEASE ATTENDING CHHATTISGARH INSTITUTE OF MEDICAL SCIENCES BILASPUR CHHATTISGARH

Hematology

Dr. Bhanu Pratap Singh Director, Professor and Head Department of Pathology, Chhattisgarh Institute of Medical Sciences Bilaspur Chhattisgarh

Dr. Nilopher Hirani* Post graduate resident Department of Pathology, Chhattisgarh Institute of Medical Sciences Bilaspur*Corresponding Author

Dr. Jyoti Porte Assistant Professor Department of Pathology, Chhattisgarh Institute of Medical Sciences Bilaspur Chhattisgarh Pin code – 495001

ABSTRACT

INTRODUCTION Sickle cell disease is an inherited hemoglobinopathy characterized by chronic hemolytic anemia, Vaso-occlusive crisis, and multiple organ complications. Hematological parameters play an important role in the assessment and management of children with sickle cell disease. **AIM** To evaluate hematological parameters in children diagnosed with sickle cell disease. **MATERIAL AND METHODS** It was a cross-sectional descriptive study conducted in the department of Pathology Chhattisgarh Institute of Medical Sciences Bilaspur. Total 50 Children diagnosed with sickle cell disease attending CIMS hospital were included in the study. Complete hemogram was done in the institutional hematology laboratory using automated hematology analyzer. Hematological parameters namely hemoglobin, total WBC count, total platelet count, PCV, MCV, MCH, MCHC and RDW were recorded and analyzed. **RESULT** Total 50 children with sickle cell disease were included in this study. Out of which 27 (54%) were male and 23 (46%) were female. 12 (24%) children were in the age group of 2-5 yrs, 25 (50%) children were in the age group of 6-10 yrs and 12 (24%) children were in the age group of 11-14 years. Our study showed that 26 (52%) children were suffering with severe anemia, 7 (14%) children were having MCV less than <70 fl, 43(86%) children were having MCV between 70-100 fl. **DISCUSSION** Our study included 50 children with sickle cell disease. Out of them 26 were suffering with severe anemia.

KEYWORDS

Sickle cell disease, Hematological profile, Hemolysis, Anemia, Hemoglobin S.

INTRODUCTION

Sickle cell anemia is a hereditary hematological disorder with profound implication for affected individuals and their families. It is characterized by the production of abnormal hemoglobin, known as hemoglobin S (HbS), due to a single point mutation in the beta-globin gene, resulting in the substitution of valine for glutamic acid at position 6[1]. This mutation leads to the polymerization of HbS under low oxygen conditions, causing red blood cells to deform into a sickle shape. These sickled cells exhibit reduced flexibility and impaired oxygen transport, leading to a variety of clinical manifestations, including Vaso-occlusive crises, chronic hemolytic anemia, and multi-organ damage[2]. Normal red blood cell has a life span of around 90-120 days, but a sickle cell only lives for about 10 to 20 days.[3]

Chhattisgarh, a state with a significant tribal population, has a notably high prevalence of sickle cell anemia. One study showed that prevalence of sickle cell trait is 10.6% and sickle cell disease is 0.66% in Chhattisgarh[4]. Children with sickle cell disease exhibit characteristic hematological abnormalities resulting from chronic hemolysis, bone marrow hyperactivity, and recurrent inflammatory state. The hematological profile often reflects the severity of the disease and varies depending on whether the child is in steady state or experiencing a Vaso-occlusive crisis. Anemia is a hallmark feature of SCD. Affected children typically present with chronic normocytic or mildly microcytic anemia, with hemoglobin commonly ranging between 6-9 gm/dl. The reduced hemoglobin concentration is primarily due to shortened red blood cell lifespan caused by ongoing hemolysis. Hematological parameters play a crucial role in the assessment and management of children with sickle cell disease. Changes in hematological parameters can indicate treatment response or the need for adjustment in management strategies.

AIM

To evaluate hematological parameters in children diagnosed with sickle cell disease.

MATERIAL AND METHODS

It was a cross-sectional descriptive study conducted in the department of Pathology CIMS Bilaspur. Ethical clearance was obtained from institutional ethical committee. Total 50 Children diagnosed with sickle cell disease attending CIMS hospital were included in the study.

INCLUSION CRITERIA

Children confirmed to be suffering with sickle cell disease by hemoglobin electrophoresis / HPLC, falling in the age group between 2yr – 14 years were enrolled for the study.

EXCLUSION CRITERIA

1) Children with other genetic disorders or chronic disorders like renal disease or liver disease

2) Patients who have received blood transfusion within the past 1 month

3)Patient/Guardian not giving consent

After getting informed consent from both children and guardian, children were enrolled for the study. Detailed clinical history, general and systemic examination was done and recorded. For all the hematological investigations 2 ml of blood sample was collected in a EDTA vial. Complete hemogram was done in the institutional hematology laboratory using automated hematology analyzer based on the principle of impedance and light scatter. Hematological parameters namely hemoglobin, Total WBC count, Total platelet count, PCV, MCV, MCH, MCHC and RDW were recorded and analyzed.

STATISTICAL ANALYSIS

All the collected data were entered in MS EXCEL sheet and analyzed by SPSS 2.0 trial version. Continuous data were represented as mean.

RESULTS

Total 50 children with sickle cell disease were included in this study. Out of which 27 (54%) were male and 23 (46%) were female. 12 (24%) children were in the age group of 2-5 yrs, 25 (50%) children were in the age group of 6-10 yrs and 12 (24%) children were in the age group of 11-14 years.

Table 1: Study of Hemoglobin

Hemoglobin	Number of children
<7 gm/dl	26
>7 gm/dl	24

Our study showed that 26 (52%) children were suffering with severe anemia. Mean hemoglobin was 6.7 gm%.

Table 2: Study of Total WBC Count

Total WBC count	Number of children
<4000/cumm	5
4000-11000/cumm	36
>11000/cumm	10

Our study showed that 5 (10%) children were having total WBC count <4000/cumm, 36 (72%) children were having TWBC count between 4000-11000/cumm and 10 (20%) children were having TWBC count >11000/cumm. Mean TWBC count was 7,217.

Table 3: Study of Total Platelet Count

Total Platelet count	Number of children
<1.5 lacs/cumm	8
1.5-4.5 lacs/cumm	42
>4.5 lacs/cumm	00

Our study showed that 8 (16%) children were having total platelet count <1.5 lacs/cumm, 42 (84%) children were having TPC between 1.5-4.5 lacs/cumm. Mean total platelet count was 2.2 lacs/cumm.

Table 4: Study of PCV

PCV	Number of children
<35%	50
>35%	00

In our study all the children were having PCV <35%. Mean PCV was 19.4%.

Table 5: Study of MCV

MCV	Number of children
<70 FL	7
70-100 FL	43
>100 fl	00

In our study we found that 7 (14%) children were having MCV less than <70 fl, 43 (86%) children were having MCV between 70-100 fl, and none were having MCV, >100 fl. Mean MCV was 82 fl.

Table 6: Study of MCH

MCH	Number of children
<27 pg	18
>27pg	32

In our study we found that 18 (36%) children were having MCH less than <27pg and 32 (64%) children were having MCH >27pg. Mean MCH was 28.2.

Table 7: Study of MCHC

MCHC	Number of children
<30 pg	7
>30 pg	43

We found that 7(14%) children were having MCHC <30 pg and 43 (86%) children were having MCHC >30 pg. Mean MCHC was 32.

Table8: Study of RDW

RDW	Number of children
<14.5	3
>14.5	47

3(6%) children were having RDW <14.5 and 47(94%) children were having RDW >14.5. Mean RDW was 21.2.

DISCUSSION

Our study included 50 children with sickle cell disease. Out of them 26 (52%) were suffering with severe anemia. In our study mean hemoglobin was 6.7 gm/dl which was similar to the study by Akodu SO et al5, Keyur Brahme et al6 but very less as compared to findings of Meshram et al7 and Muley P et a[18]. This finding can be attributed to chronic hemolysis, reduced RBC life span and also may be due to episodic bone marrow suppression. Mean total WBC count was 7,217/cumm, which was lower as compared to Muley P et al8 and Meshram et al7. Majority of children were having WBC count within normal range, it reflects effective bone marrow function, balanced immune regulation and absence of acute infection, inflammation, or physiological stress at the time of testing. In our study mean total platelet count was 2.2 lacs/cumm which was almost near to the findings by Meshram et al7 and Muley P et a[18] but lower as compared to the findings by Keyur Brahme et al6. A normal platelet count suggests relative clinical stability, absence of acute Vaso-occlusive events. Maintenance of a normal platelet count in children with sickle cell disease may reflect preserved or partially preserved splenic function, effective disease management, and the absence of functional asplenia-related thrombocytosis. Mean PCV was 19.4% which was lower as compared to the findings by Keyur Brahme et al6 and Muley P et a[18]. Mean MCV in our study was 82 fl which was similar to the study by Keyur Brahme et al6. but higher as compared to the study by Akodu SO5. and Meshram et al7. A normal MCV in majority of the patients in this study indicates that sickle cell disease is primarily a hemoglobinopathy rather than a disorder of red cell size or hemoglobin synthesis. A child with sickle cell disease should be investigated for nutritional deficiencies if he has a low MCV. Mean MCH in our study was 28.2, which as higher as compared to the findings by Meshram et al7 and Akodu SO et al5. Mean MCHC in our study was 32, which was similar to the findings of Akodu SO et al5, higher than the findings by Keyur Brahme et al6, and lower than the findings by Meshram et a[17]. Slight deviations in MCH and MCHC correlate with the degree of hemoglobinopathy and concurrent micronutrient deficiencies. Majority were having normocytic RBCs with elevated RDW. It aligns with chronic hemolysis and dynamic erythropoiesis.

CONCLUSION

This study highlights the characteristic hematological profile of children with sickle cell disease. Severe anemia in children with sickle cell disease reflects the complex interplay of hemolysis, marrow response, infections, and nutritional factors. Alteration in key parameters including hemoglobin levels, PCV and RDW underscore the persistent hematologic stress associated with the disease, even during steady state. Understanding these hematological patterns is essential for early identification of disease complications, monitoring therapeutic response, and guiding clinical management. Regular hematological assessment remains a cornerstone in the comprehensive care of children with sickle cell disease, aiding in timely intervention and improved outcomes.

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