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# PREVALENCE OF SICKLE CELL DISEASES IN TRIBAL PATIENTS BY USING HIGH PERFORMANCE LIQUID CHROMATOGRAPHY

Pathology

**Dr. Jitendra Kumar Singh*** Junior Resident / Department of Pathology / R.I.M.S./ Ranchi University , India *Corresponding Author

**Dr. Sunil Kumar Mahto** Associate Professor / Department of Pathology / R.I.M.S./ Ranchi University , India

**Dr. Devendu Bose** Junior Resident / Department of Pathology / R.I.M.S./ Ranchi University , India

**Dr. Shweta Kumari** Junior Resident / Department of Otolaryngology / R.I.M.S./ Ranchi University , India

## ABSTRACT

The inherited disorders of blood include sickle cell diseases as one of the major public health problem in India. The Hemoglobin sickle (HbS) syndrome is frequently seen in various part of Jharkhand. It includes- Sickle cell disease, sickle cell trait, sickle cell β thalassemia. The tribes of Jharkhand consist of 32 tribes inhabiting the Jharkhand state in India.

Automated cation-exchange High Performance Liquid Chromatography (HPLC) has emerged as an excellent screening tool for diagnosing these abnormal hemoglobin/thalassemic states.

The present study is carried out to correctly diagnose the sickle cell diseases by performing HPLC and family studies if possible, and to generate data so that health care resources can be successfully planned and targeted at them.

The present study was done at the Department of Pathology, Rajendra Institute of medical sciences, Ranchi between July,2018 to September 2019.

Majority (41.7%) of cases were found below the age group of 10 years. Most of them (43.3%) were found to have sickle cell trait. Pallor was the commonest feature among all the age groups, majority having Hb level in between 7-9.9 gm %. 20.0% of cases showed consanguinity.

HPLC is reliable, reproducible, and in many cases superior to conventional hemoglobin electrophoresis for the detection and identification of hemoglobin variants.

## KEYWORDS

## I.INTRODUCTION

The sickling disorders like HbSS, HbSD, HbSE, HbS/β- thalassemias are all clinically significant, as these combinations present with different manifestations and degrees of severity, making precise identification important.[1] There is wide variation in the longevity and clinical pattern of sickle cell disorders in different communities. The recognized factors that tend to affect the variability include age, concomitant presence of thalassemia gene, high level of fetal hemoglobin, presence of glucose-6-phosphate dehydrogenase deficiency and environmental factors like hypoxia, dehydration, infection, acidosis, extreme of temperature and emotional stress.[2] Automated cation-exchange High Performance Liquid Chromatography (HPLC) has emerged as an excellent screening tool for diagnosing these abnormal hemoglobin/thalassemic states.[1] Hemoglobin fraction analysis by cation-exchange HPLC has the advantage of quantifying HbF and HbA2 along with hemoglobin variant screening in a single, highly reproducible system, making it an excellent technology to screen for hemoglobin variants and hemoglobinopathies along with the thalassemias.[3] The definite identification of disorders of hemoglobin synthesis can be achieved only by DNA analysis but in Indian scenario, family studies on HPLC might be useful as there is paucity of funds and facilities for DNA analysis are not readily available.[1] Family study is an equally efficacious and cost effective tools. The tribes of Jharkhand consist of 32 tribes inhabiting the Jharkhand state in India. The tribes in Jharkhand were originally classified on the basis of their cultural types by the Indian anthropologist, Lalita Prasad Vidyarthi.

**Her classification was as follows:**

1. Hunter-gatherer type — Birhor, Korwa, Hill Kharia
2. Shifting Agriculture — Sauria, Paharia
3. Simple artisans — Mahli, Lohra, Karmali, Chik Baraik
4. Settled agriculturists — Santhal, Munda, Oraon, Ho, Bhumij, etc.

The retention time on HPLC is reliable, reproducible, and in many cases superior to conventional hemoglobin electrophoresis for the detection and identification of hemoglobin variants.[3]

## II.MATERIAL AND METHODS

A total of 60 cases of Hemoglobin sickle (HbS) syndrome were included for clinical and hematological features.

**Study Design:** Cross-sectional Descriptive study

**Study Location**: Department of Pathology at Rajendra Institute of Medical Sciences, Ranchi

**Study Duration:** July 2018 to September 2019

**Sample size:** 60 patients.

**Inclusion Criteria:**

All patients of anemia showing sickling test positive.
Patients with clinical presentation of hemolytic anemia.
Patients with hepato-splenomegaly.
Family members of these patients.

**Exclusion Criteria:**

Patients who had received blood transfusion in last 6 month were excluded from study.

## III.Observations and Results

**Table 1 : Showing distribution of Study subjects according to impression.**

| Impression | Frequency | Percent |
|---|---|---|
| sickle beta thalassemia | 12 | 20.0% |
| sickle cell disease | 21 | 35.0% |
| sickle cell trait | 26 | 43.3% |
| sickle E disease | 1 | 1.7% |
| Total | 60 | 100.0% |

Out of 60 study subjects, 26(43.3%) were found sickle cell trait followed by 21 (35%) having sickle cell disease and 12 (20%) having sickle beta thalassemia whereas 1(1.7%) was found sickle E disease.

**Table 2: Showing distribution of Study subjects according to Age group**

| Age Group | Frequency | Percent |
|---|---|---|
| <10yrs | 25 | 41.7% |
| 10-20yrs | 22 | 36.7% |
| 20-30yrs | 11 | 18.3% |
| >30yrs | 2 | 3.3% |
| Total | 60 | 100.0% |

Out of 60 study subjects Majority 25(41.7%) found below the age group of 10 years, followed by 22(36.7%) in age group between 10-20 years and 11(18.3%) were found to be between 20-30years of age group, whereas remaining 2(3.3%) were found to be age group above 30 years.

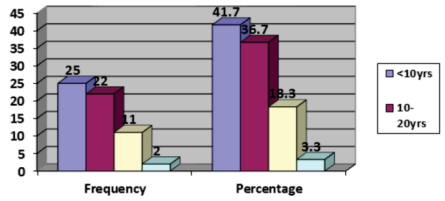


**Figure1: Showing distribution of Study subjects according to Age group.**

**Table 3 : Showing distribution of Study subjects according to presence of consanguinity.**

| Consanguinity | Frequency | Percent |
|---|---|---|
| NO | 48 | 80% |
| YES | 12 | 20% |
| Total | 60 | 100.0% |

Out of 60 study subject, 12 (20%) were showing presence of consanguinity while 48 (80%) were showing absence of consanguinity.

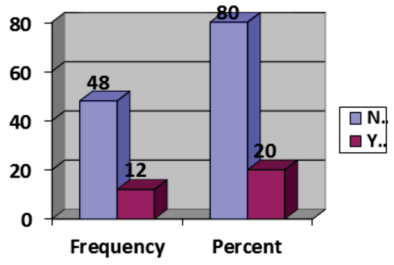


**Figure 2 : Showing distribution of Study subjects according to presence of consanguinity.**

**Table 4: Showing distribution of Study subjects according to Hemoglobin level.**

| Hb level (gm%) | Frequency | Percent |
|---|---|---|
| <7 | 19 | 31.7% |
| 7-9.9 | 32 | 53.3% |
| 10-11.9 | 8 | 13.3% |
| >12 | 1 | 1.7% |
| Total | 60 | 100.0% |

Out of 60 study subjects, Hb level of majority i.e, 32(53.3%) found in between 7-9.9 gm % followed by 19(31.7%) below 7 gm % and 8(13.3%) were found in between 10-11.9 gm %, whereas remaining 1 (1.7%) was found above 12 gm %.

**Table 5: Showing distribution of Study subjects according to Clinical Features**

| Clinical Features | Number of Cases |
|---|---|
| Pallor | 60 |
| Pain abdomen | 16 |
| Vaso occlusive crisis. | 2 |
| Leg ulcer | 5 |
| Splenomegaly | 35 |

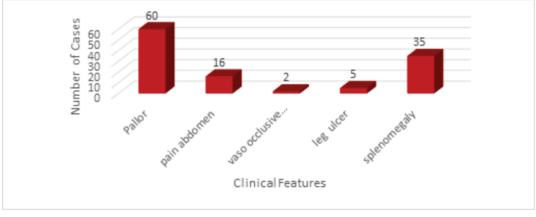


**Figure 4: Showing distribution of Study subjects according to Clinical Features**

## IV.DISCUSSION

The present study entitled " Prevalence of Sickle Cell Diseases in tribal patients by using High Performance Liquid Chromatography" included 60 cases of sickling disorder after fulfilling inclusion and exclusion criteria.

In the present study, out of 60 study subjects Majority 25(41.7%) found below the age group of 10 years, followed by 22(36.7%) in age group between 10-20 years and 11(18.3%) were found to be between 20-30years of age group, whereas remaining 2(3.3%) were found to be age group above 30 years. Tyagi et al[4] in 2003 reported 16 cases of Sβ thalassemia out of 47 cases of sickling disorders and the average age of presentation was 14.2 years.

In the present study of the total 60 sickling disorders (SCA, SCT, Sβ thal, sickle E disease), 26(43.3%) were found sickle cell trait followed by 21 (35%) having sickle cell disease and 12 (20%) having sickle beta thalassemia whereas 1(1.7%) was found sickle E disease.

Pallor was the most common feature among all the sickling disorders followed by splenomegaly(58.3%), jaundice(43.3%) and pain abdomen(26.7%).

In the present study splenomegaly has been seen in 58.3% of cases. R.S Balgin (2010) et al[5] found splenomegaly in 17.1% of cases. Splenomegaly is higher in patients with Sβ thalassemia than SS disease In the present study, there were 12 (20%) cases with history of consanguineous marriage. Consanguinity is commonly practiced in Muslims but it is also common in rural tribals of central India. Patel DK[6] had stated that in India the practice of consanguineous marriages are very common, and since β- thalassemia and hemoglobin S (HbS) are relatively frequent in India, compound heterozygosity for these two disorders (βS/β- thalassemia), usually expressed in a severe type of disease, is not rare. Tariq HA[7] narrated that, the main factor, which may increase or decrease the risk of having an affected child, is the parents relationship.

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