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Research Article

STUDY OF CLINICO-HEMATOLOGICAL PROFILE OF HEMOGLOBINOPATHIES AT TERTIARY CARE CENTRE

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ABSTRACT

Background: Inherited hemoglobin disorders constitutes major bulk of genetic diseases in India. Inherited hemoglobin disorders include structural hemoglobin variants and thalassemia. Structural hemoglobin variants mostly result from amino acid substitution in either α or β chains.The cumulative gene frequency of hemoglobinopathies in India is 4.2%.
Methods: A total of 150 patients were examined. We have examined all patients with anemia or suspected beta thalassemia. The patient groups were evaluated according to the clinical presentation.
Results: Sickle cell trait was the most common hemoglobinopathy detected. Occurrence of hemoglobinopathies was highest in Buddhas. Pallor was the most common symptom.
Conclusions: Sickle cell trait was the most common hemoglobinopathy detected.
Occurrence of hemoglobinopathies was highest in Buddhas, followed by Muslim and Banjaras. Most common affected ethnic group was Buddha followed by Banjaras in sickle cell syndromes. Pallor was the most common symptom in present study [60 %], followed by jaundice [14.70 %]. Vaso-occlusive crisis [abdominal pain, joint pain] was more common in sickle /beta thalassemia than sickle cell anaemia, while jaundice was more common in sickle cell anaemia than sickle /beta thalassemia. Jaundice was found to be less common in beta-thalassemia major. Splenomegaly was more common in beta thalassemia major than sickle cell anemia. Hplc was found to be less labour intensive, rapid and more reliable for quantification of hemoglobin variants. Family studies are required to confirm the diagnosis.

INTRODUCTION

Inherited hemoglobin disorders constitutes major bulk of genetic diseases in India. Inherited hemoglobin disorders include structural hemoglobin variants and thalassemia. Structural hemoglobin variants mostly result from amino acid substitution in either α or β chains.

It has been estimated that 7% of the world populations are carriers of such disorders and that 300000-400000 babies with severe forms of these diseases are born each year.[1]Around 1.1% of couples worldwide are at risk for having children with a hemoglobin disorder and 2.7 per 1000 conceptions are affected. Over 9 million carriers become pregnant annually. The risk that their partner is also a carrier ranges from 0.1-40% (global average 14%).[2]

The cumulative gene frequency of hemoglobinopathies in India is 4.2%. With a population of over one billion and a birth rate of 28 per thousand, there are over 42 million carriers and over 12,000 infants are born each year with a major and clinical significant hemoglobinopathy.[3] Automated cation exchange high performance liquid chromatography (hplc) has emerged as an excellent screening tool for diagnosing these thalassaemic states.[1] Haemoglobin fraction analysis by cation-exchange hplc has the advantage of quantifying HbF and HbA_2 along with haemoglobin variant screening in a single, highly reproducible system, making it an excellent technology to screen for haemoglobin variants and haemoglobinopathies along with the thalassaemias.

The simplicity of the automated system makes this an ideal methodology for the routine clinical laboratory. Exact diagnosis of these diseases is of paramount importance in therapy and prevention of genetic transmission. This study was carried out to study clinico-hematological profile of hemoglobinopathies.

METHODS

A total of 150 clinically and haematologically suspected cases of haemolytic anaemia during the period of October 2013 to October 2015 were selected.

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Two ml of intravenous blood sample was collected from all cases after obtaining informed consent using ethylene diamine tetra acetic acid (EDTA) as anticoagulant.

All patients with normal Hemoglobin, but having thalassemic blood indices on coulter.[i.e MCV<80,MCH<27], all patients of anemia (OPD and Indoor) showing sickling test positive, patients presenting with hepatosplenomegaly. patients with clinical suspicion of haemolytic anemia, family members of these patients were used as inclusion criteria.

Patient received blood transfusions in last three months were excluded from study.

Table 1 Analysate identification windows[4]

Retention time (minutes)	Band (minutes)	Window (minutes)	Range
F	1.15	0.15	1.00-1.30
P2	1.45	0.15	1.30-1.60
P3	1.75	0.15	1.60-1.90
A0	2.60	0.40	2.20-3.30
A2	3.83	0.15	3.68-3.98
D-window	4.05	0.15	3.98-4.12
S-window	4.27	0.15	4.12-4.42
C-window	5.03	0.15	4.88-5.18

PS, CBC including RBC Indices, Reticulocyte count, sickling test and if required, bone marrow was done. Samples were run on HPLC machine Bio-Rad variant-II and hemoglobin graph was obtained and diagnosis of hemoglobinopathies was confirmed using values of different hemoglobin fractions and retention times (Table 1).

Family study: Family studies of cases was carried out wherever possible, to confirm the diagnosis as family study is effective for centers which do not have facility for genetic analysis. Mother, father, siblings, son and daughter of patient were studied.

Family study was advised, but may not be completed due to:

- Distance
- Remarriage
- Death
- Poor Socioeconomic status
- Reluctance in fathers
- Lack of awareness
- Misconceptions
- Alcoholism in fathers
- Psychological reasons.

RESULTS

In the present study, total 150 suspected cases of hemoglobin disorders were studied by HPLC in Department of Pathology in Tertiary Care Hospital from October 2013 to October 2015.

Table No 2 Diagnosis of cases by HPLC in present study

No.	Diagnosis	CASES
1.	Sickle Cell Trait (SCT)	43
2.	Sickle Cell Disease (SCD)	9
3.	Sickle Cell - Beta Thalassemia (SBT)	18
4.	Beta Thalassemia Trait (BTT)	32
7.	Beta Thalassemia Major (BTM)	16
8.	HbE - Beta Thalassemia	1
9.	Hb Lepore Trait	1
10.	Normal Pattern	30

120 cases were diagnosed as Hemoglobin disorders out of 150 cases by HPLC, So cases were classified as

Group-A Sickle Cell Trait (SCT)
Group-B Sickle Cell Disease (SCD)
Group-C Sickle Cell - Beta Thalassemia (SBT)
Group-D Beta Thalassemia Trait (BTT)
Group-E Beta Thalassemia Major (BTM)
Group-F HbE - Beta Thalassemia
Group-G Hb Lepore Trait

In the present study HPLC was considered as standard method. A total of 30 cases suspected to be hemoglobinopathies were found to be normal by HPLC, these 30 cases were taken as normal control group.

Table No 3 Groupwise Classification of Hemoglobin disorders in present study

Group	Diagnosis	No. of cases
A	Sickle Cell Trait (SCT)	43 [35.83%]
B	Sickle Cell Disease (SCD)	9 [7.5%]
C	Sickle Cell - Beta Thalassemia (SBT)	18 [15%]
D	Beta Thalassemia Trait (BTT)	32 [26.66%]
E	Beta Thalassemia Major (BTM)	16 [13.33%]
F	Hemoglobin E-Beta Thalassemia	1 [0.83%]
G	Hb Lepore Trait	1 [0.83%]
Total cases of Hemoglobin disorders		120

Table No 4 Age wise distribution of cases among all the groups in present study

Age (Yrs)	Gr-A n=43	Gr-B n=9	Gr-C n=18	Gr-D n=32	Gr-E n=16	Gr-F n=1	Gr-G n=1
0 to 10	12 (8%)	1 (0.66%)	4 (2.66%)	2 (1.33%)	15 (10%)	0 (0%)	0 (0%)
11 to 20	10 (6.6%)	6 (4%)	7 (4.66%)	0 (0%)	1 (0.66%)	0 (0%)	0 (0%)
21 to 30	15 (10%)	2 (1.33%)	5 (3.33%)	16 (10.66%)	0 (0%)	1 (0.66%)	0 (0%)
31 to 40	3 (2%)	0 (0%)	2 (1.33%)	11 (7.33%)	0 (0%)	0 (0%)	0 (0%)
41 to 50	3 (2%)	0 (0%)	0 (0.0%)	2 (1.33%)	0 (0%)	0 (0%)	1 (0.66%)
51 to 60	0 (0%)	0 (0%)	0 (0.0%)	1 (0.66%)	0 (0%)	0 (0%)	0 (0%)
61 to 70	0 (0%)	0 (0%)	0 (0.0%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)

In Group-A i.e. Sickle cell trait

Maximum cases were in 21 to 30 years age group.

In Group-B i.e. Sickle cell disease

Maximum cases were in 11 to 20 years age group.

In Group-C i.e. Sickle-beta thalassemia

Maximum cases were in 11 to 20 years age group.

In Group-D i.e. Beta Thal trait

Maximum cases were in 21 to 30 years age group.

In Group-E i.e. Beta Thal Major

Maximum cases was found in 0 to 10 age group.

In Group-F i.e. HbE Beta Thal

1 case was found in 21 to 30 years.

In Group-G i.e. Hemoglobin Lepore Trait

1case was found in 41 to 50 years.

Table No 5 Sex Wise Distribution of cases among all groups in present study

Group	Male	Female	Total
Gr-A	17	26	43
Gr-B	4	5	9
Gr-C	12	6	18
Gr-D	12	20	32
Gr-E	10	6	16
Gr-F	1	0	1
Gr-G	1	0	1

In Group-A i.e. Sickle cell trait

No. of Male patient 17 and No. of Female patient 26

In Group-B i.e. Sickle cell disease

No .of Male patient 4 and No.of Female patient 5.

In Group-C i.e. Sickle-beta thalassemia

No. of Male patient 12 and No. of Female patient 6.

In Group-D i.e. Beta Thal Trait

No. of Male patient 12 and No. of Female patient 20.

In Group-E i.e. Beta Thal Major

No. of Male patient 10 and No. of Female patient 6.

In Group-F i.e. HemoglobinE-beta Thalassemia

No. of Male patient 1

In Group-G i.e. Hemoglobin Lepore Trait.

No. of Male patient 1

Table No 6 Caste wise distribution of cases among all groups in present study

Caste	GROUP							Total	%
	A	B	C	D	E	F	G		
Buddha	22	5	11	14	7	0	0	59	39.3
Muslim	3	1	2	8	6	1	0	21	14
Banjara	9	2	4	2	2	0	1	20	13.3
Mang	0	1	0	2	1	0	0	4	2.66
Mali	2	0	0	1	0	0	0	3	2
Bhil	4	0	0	0	0	0	0	4	2.66
Maratha	1	0	0	1	0	0	0	2	1.33
Dhangar	1	0	1	2	0	0	0	4	2.66
Nepali	1	0	0	1	0	0	0	2	1.33
Chambhar	0	0	0	1	0	0	0	1	0.66

Buddha (39.3%) was the most common ethnic background among all the groups followed by Muslims (14%) then Banjara (13.3).

Table No 7 Clinical Presentation of cases among all Groups in present study

Clinical features	Group							Total	% out of total 150 cases
	A [n=43]	B [n=9]	C [n=18]	D [n=32]	E [n=16]	F [n=1]	G [n=1]		
Pallor	14	9	18	16	16	1	0	74	49.33
Jaundice	3	9	5	1	1	1	0	20	13.33
Joint pain	2	2	7	0	0	0	0	11	7.33
Pain in Abdomen	0	1	11	0	3	0	0	15	10

Majority of the cases presented clinically with Pallor 49.33%, next common presentation was jaundice. Other clinical features were pain in abdomen and joint pain

Table No 8 Distribution of cases with Splenomegaly among all groups in present study

Age group (Yrs)	Group B (SCD)	Group C (SBT)	Group D (BTT)	Group E (BTM)	Group F (HbE β)	Total [n=27]	Per. (%)
0-10	0	4	0	14	0	18	66.66%
11-20	1*	4	0	1	0	6	22.22%
21-30	0	1	1**	0	1	3	11.11%
31-40	0	0	0	0	0	0	0%

Splenomegaly was detected in one case out of nine cases of Group B.

Out of 18 cases of Group C, 9 presented with Splenomegaly.
In Group D, out of 32 cases Splenomegaly was detected in one case. In the Group E, splenomegaly was found in 15 patients [93.75%].

In the Group F, splenomegaly was found in one case.
In Group A and Group G, No Splenomegaly detected.

Out of 120 cases 27 [22.5 %] cases presented with splenomegaly. Out of 27 cases, 18 cases were found in 0-10 year's age group, followed by 6 cases in 11-20 years age group. So the maximum cases of splenomegaly were found in first decade.

Table No.9 Age wise Distribution of Clinical presentation of Group-B (SCD)*

	Group-B (SCD)* n=9			
Age Group	Pallor	Jaundice	Joint pain	Abdominal pain
0-10	1 [11.11%]	0 [0%]	0 [0%]	0 [0%]
11-20	6 [66%]	2 [22.22%]	0 [0%]	1 [11.11%]
21-30	2 (22.22%)	2 (22.22%)	1 (11.11%)	1 (11.11%)

[SCD*-Sickle Cell Disease]
Maximum cases were present in second decade, pallor was most common symptom, followed by jaundice.

Table No 10 Age wise Distribution of Clinical presentation of Group-c (SBT)*

	Group-C (SBT)* n=18			
Age Group	Pallor	Jaundice	Joint pain	Abdominal pain
0-10	4 (22.2%)	2 (11.11%)	2 (11.11%)	1 (5.55%)
11-20	7 (38.88%)	5 (27.77%)	2 (11.11%)	3 (16.66%)
21-30	5 (27.77%)	3 (16.66%)	3 (16.66%)	4 (22.22%)
31-40	2 [11.11%]	0 (0%)	0 (0%)	0 (0%)

[SBT*- Sickle cell-Beta Thalassemia]
Maximum patients presented in second decade, with pallor [38.88%] followed by jaundice.

Table No 11 Age wise Distribution of Clinical presentation of Group-E (BTM)*

	Group-E (BTM)* n=16			
Age Group	Pallor	Jaundice	Joint pain	Abdominal pain
0-10	15 (93.75%)	2 (12.5%)	0 (0%)	3 (18.75%)
11-20	1 (6.25%)	0 (0%)	0 (0%)	0 (0%)

[*BTM-Beta Thalassemia Major]

15 cases presented in first decade, Pallor is most common symptom, followed by abdominal pain. Jaundice was less common than other Hemoglobin disorders [such as SCD, Sickle/Beta thal]. 1 case present in second decade presented with anemia.

Table No 12 Group wise distribution of Mean Hematological Parameters observed in all 150 cases

Hematological Parameters	Group						
	A	B	C	D	E	F	G
Hbg/dl	10.8	5.95	6.56	10.1	4.1	3.4	12.1
RBC million/mm^3	4.42	2.26	2.79	5.06	2.31	2.12	5.65
MCV fl	80.7	82.8	75.5	66.2	64.4	59	70.3
MCH pg	24.3	26.08	23.4	19.9	17.9	16	21.4
MCHC g/dl	30.0	31.4	31.1	30	27.7	27.1	30.4

Group A - Cases presented with mild anaemia.
Group B- Cases presented with moderate to severe anaemia, normal MCV, MCH, MCHC.
Group C- Cases presented with moderate to severe anaemia, but reduced MCV, MCH.
Group D- Cases presented with mild anaemia, normal RBC Count, Reduced MCV, MCH.
Group E- Cases presented with severe anaemia, Reduced RBC Count, Reduced MCV,MCH, and MCHC.
Group F- Case presented with severe anaemia with reduced mMCV, MCH, and MCHC.
Group G- Cases presented with normal Hemoglobin level, but raised RBC count, with reduced MCV, MCH.

DISCUSSION

The present study entitled-"Study of Clinico-Hematological Profile of Hemoglobinopathies At Tertiary Care Centre" was carried out in department of pathology in a tertiary care hospital from October 2013 to 0ctober 2015.

Inherited Hemoglobin Disorders constitutes major bulk of genetic diseases in India. Inherited Hemoglobin Disorders include structural Hemoglobin variants and thalassemia. Structural Hemoglobin variants mostly result from amino acid substitution in either α or β chains. It has been estimated that 7% of the world populations are carriers of such disorders and that 300000-400000 babies with severe forms of these diseases are born each year.[1] Around 1.1% of couples worldwide are at risk for having children with a Hemoglobin disorder and 2.7 per 1000 conceptions are affected. Over 9 million carriers become pregnant annually. The risk that their partner is also a carrier ranges from 0.1–40% (global average 14%).[2]

The cumulative gene frequency of Hemoglobinopathies in India is 4.2%. With a population of over one billion and a birth rate of 28 per thousand, there are over 42 million carriers and over 12,000 infants are born each year with a major and clinical significant Hemoglobinopathy.[3]

Table NO.13 Diagnosis of cases by HPLC in present study

No.	Diagnosis	CASES
1.	Sickle Cell Trait (SCT)	43
2.	Sickle Cell Disease (SCD)	9
3.	Sickle Cell - Beta Thalassemia (SBT)	18
4.	Beta Thalassemia Trait (BTT)	32
7.	Beta Thalassemia Major (BTM)	16
8.	HbE - Beta Thalassemia	1
9.	Hb Lepore Trait	1
10.	Normal Pattern	30

Diagnosis of the Cases by HPLC

120 cases were diagnosed as Hemoglobin disorders out of 150 cases by HPLC, So cases were classified as:

Group-A Sickle Cell Trait (SCT)
Group-B Sickle Cell Disease (SCD)
Group-C Sickle Cell - Beta Thalassemia (SBT)
Group-D Beta Thalassemia Trait (BTT)
Group-E Beta Thalassemia Major (BTM)
Group-F HbE - Beta Thalassemia
Group-G Hb Lepore Trait

In the present study HPLC was considered as standard method. A total of 30 cases suspected to be hemoglobinopathies were found to be normal by HPLC, these 30 cases were taken as normal control group.

Study of Hemoglobinopathies

In present study, **Sickle cell trait was most common hemoglobinopathy detected**. Next common Hemoglobinopathy was Beta-Thalassemia trait, followed by Sickle-Beta thalassemia, Beta-Thalassemia Major, Sickle cell anaemia. Hb- Lepore trait [1case], HbE-beta Thalassemia [1case] were rare Hemoglobinopathies detected.

Table No 14 Study of Hemoglobinopathies

Other studies	Place of Study	Prevalent Hemoglobinopathy
Ambekar S.S *et al*[5] (2001)	Maharashtra	Beta-Thalassemia trait
Sachdev R *et al* [6] (2010)	Northen India	Beta-Thalassemia trait
Rao S *et al*[7] (2010)	New Delhi	Beta-Thalassemia trait
Goswami BK *et al*[8](2010)	North Bengal (Hospital based)	HbE-trait
C.Vani and Soni M.[9] (2011)	South India	Beta-Thalassemia trait
Patel AG *et al* [10] (2012)	South Gujrath	Beta-Thalassemia trait
Baruah MK *et al* [11] (2012)	Upper Assam Region (North-Estern India).	HbE-trait
Mandal PK *et al* [12] (2013)	West Bengal	Beta-Thalassemia trait
Shrivastava A *et al*[13] (2013)	Western India	Beta-Thalassemia trait
Mondal SK *et al*[14] (2014)	West Bengal	Beta-Thalassemia trait

From above mention studies, Beta-Thalassemia trait is most common hemoglobinopathy in India. In present study, Sickle cell trait being most common hemoglobinopathy detected as present study was Hospital based, not Community based. During period of study, large numbers of solubility positive samples from peripheral health centers were received.

Ethnicity

In the present study Occurrence of hemoglobinopathies was highest in Buddhas (39.3 %), followed by Muslim (14 %) and Banjara (13.3%).

Most common affected ethnic group was Buddha followed by Muslim in Beta Thalassemias [Group D,E].

Most common affected Ethnic group was Buddha followed by Banjara in Sickle cell Syndromes [Group A, B, C].

One case of Hb-Lepore trait [Group G] was noted in Banjara.
One case of HbE-Beta Thalassemia [Group F] was noted in Muslim.

Other Studies

Bhatia and Rao 1987[15] found prevalence of sickle cell disorder is very high amongst tribal and scheduled caste population group where carrier frequencies range between 5-40% and more.

Ambekar S.S. *et al*[5] 2001reported maximum cases of beta thalassemia amongst Navbuddhas.

Kamble M, Chaturvedi P[16] observed maximum incidence of sickle Hemoglobinopathy in Mahars (70%) followed by Kunbhis (8%) and Telis (6%).

Dangi *et al*[17][2010], found Muslims, Sindhis and Tribal population the major carriers of sickle gene.

Clinical Features

Pallor

Was most common symptom in present study [49.33 %], followed by jaundice.

Most of the patients in Group A [SCT] were asymptomatic. They were included in present study 1] Due to positive solubility 2] As part of Family study.

In Group B [SCA], Pallor was most common symptom followed by jaundice.

In Group C [Sickle-Beta Thal], Pallor was most common symptom followed by Abdominal pain.

Most of patients in Group D [BTT] were asymptomatic. They were included in present study 1] As part of Family study. 2] To rule out Beta- Thalassemia as cause of microcytic anaemia.3] Microcytic anaemia not responding to iron therapy.
In Group E [BTM], Pallor was most common symptom followed by Abdominal pain. Jaundice was less common than other Hemoglobinopathies [Sickle cell anaemia, Sickle-beta Thalassemia].

In Group F [HbE-Beta-Thal], case presented with Pallor, jaundice.

In Group G [Hb-Lepore Trait], Case was asymptomatic. Solubility was false positive.

Other Studies

Tyagi *et al*[18]in 2003 reported 47 cases of sickling disorders. Twenty of the 47 patients (42.6%) were clinically asymptomatic. The majority were either picked up during family studies or during investigation of unrelated symptoms, such as pregnancy, mild anaemia, hypertension or diabetes mellitus.

Tyagi *et al* also found that VOC (81.3%) was the most common clinical feature followed by pallor (56.3%) in Sickle Beta-Thalassemia. Pallor (62.5%), jaundice (75%), painful crisis (62.5%) and leg ulceration (25%) were more frequent in sickle cell disease than in double heterozygous Sβ Thal patients.

RS Balgir *et al*[19] in Apr-2010 reported 137 cases of sickling disorders. R.S. Balgir (2010) observed that pallor was more frequently a clinical presentation in Sβ Thalassemia cases (58.6%) followed by VOC (31%).

Panigrahi I *et al*[20] (2005) in their study on HbE-β Thal patients found pallor as the commonest feature(100%), splenomegaly in 74% cases, hepatomegaly in 65% cases, jaundice in 57% cases.
Shah SJ *et al*[21] (2012) studied 'A profile of cases of Hemoglobinopathies at a medical college' reported-

- 35 out of 35 cases with pallor (100%),
- 8 cases with icterus (22.9%)

Splenomegaly

Splenomegaly was detected in one case out of ninecases of Sickle Cell Disease (Group B)

Out of 18 cases of Group C [Sickle-Beta Thalassemia] , 9 presented with Splenomegaly. Splenomegaly was more common in sickle-Beta thalassemia than sickle cell Anemia [0 cases with splenomegaly], also persists longer than sickle cell anemia [cases in second and third decade]

In Group D [Beta Thalassemia Trait] out of 32 cases Splenomegaly was detected in one case.

In the Group E [Beta Thalassemia major]- splenomegaly was found in 15 patients [93.75%].

In the Group F [HbE beta Thalassemia]-splenomegaly was found in one case.

In Group A [Sickle Cell Trait] ,Group G [Hb Lepore Trait] - No Splenomegaly detected.

Other Studies

R.S.Balgir *et al*[19] (2010) found splenomegaly in 17.1% of cases. Splenomegaly is higher in patients with Sβ Thalassemia than SS disease.

Patel DK[22] stated that persistent gross splenic enlargement is peculiarity of Indian sickle disorder patients.

Tyagi *et al*[18] (2003) in their study on sickle cell syndrome found that the frequency of splenomegaly was significantly higher (75%) in patients with Sβ Thal than Sickle cell disease.
Panigrahi I *et al*[20] (2005) in their study on HbE-β Thal patients found splenomegaly in 74% cases.

Shah SJ *et al*[21] (2012) reported

- 31 cases with splenomegaly (88.6%),
- 25 cases with hepatomegaly (71.4%)

Relevant Haematological Parameters

Table No 15 Relevant Haematological Parameters in Group A [SCT*]

Studies	Hb g/dl	RBC Million/mm3	MCV fl	MCH Pg	MCHC g/dl
Rao S *et al*[7][2008]	11.6	4.45	84.3	26.7	31.7
Shrivastav *et al*[13][2013]	9.89	4.49	70.27	22.2	31.4
Mondal *et al*[14] [2014]	10.8	3.8	85.2	27.4	32.4
Present study [2015]	10.8	4.42	80.7	24.3	30

[SCT*-Sickle Cell Trait]

Cases presented with mild anaemia, with near normal CBC parameters.

Haematological parameters of present study are consistent with Rao S *et al* study.

Table No.16 Relevant Haematological Parameters in Group B [SCD*]

Studies	Hb g/dl	RBC Million/mm3	MCV fl	MCH pg	MCHC g/dl
Rao S *et al*[7][2008]	8.3	2.8	90.5	29.5	32.7
Shrivastav *et al*[13][2013]	7.46	3.44	75.7	25.6	32.9
Mondal *et al*[14][2014]	7.8	2.7	91.4	30.2	33.3
Present study [2015]	5.95	2.26	82.8	26.08	31.4

[SCD*-Sickle Cell Disease]

Cases presented with moderate to severe anaemia, normalMCV, MCH, MCHC. Hemoglobin level is low as compared to other studies in present study, as patient who has received blood transfusion in last three months was excluded from study.

Table No. 17 Relevant Haematological Parameters in Group C [Sickle-Beta Thalassemia]

Studies	Hb g/dl	RBC Million/mm3	MCV fl	MCH pg	MCHC g/dl
Rao S *et al* [7] [2008]	7.6	3.49	75.2	21.8	29.2
Shrivastav *et al*[13] [2013]	7.91	3.62	70.28	22.5	32.1
Mondal *et al* [14] [2014]	7.8	3.2	76.6	22.3	30.2
Present study [2015]	6.56	2.79	75.5	23.4	31.1

Cases presented with moderate to severe anaemia, but reduced MCV, MCH. Haematological parameters of present study are consistent with Rao S *et al* study.

Table No 18 Relevant Haematological Parameters in Group D [Beta-Thalassemia trait]

Studies	Hb g/dl	RBC Million/mm3	MCV fl	MCH Pg	MCHC g/dl
Rao S *et al*[7] [2008]	10.3	5.06	68.6	20.5	28.3
Shrivastav *et al*.[13] [2013]	10.4	5.38	62.1	19.4	30.3
Mondal *et al* [14] [2014]	9.7	3.8	70.4	21	27.8
Present study [2015]	10.1	5.06	66.2	19.9	30

Cases presented with mild anaemia, with normal RBC Count, Reduced MCV, MCH. Haematological parameters of present study are consistent with Rao S *et al* study.

Table No 19 Relevant Haematological Parameters in Group E [Beta-Thalassemia Major]

Studies	Hb g/dl	RBC Million/mm3	MCV fl	MCH Pg	MCHC g/dl
Rao S *et al* [7] [2008]	5.4	2.41	74.9	23.3	31.1
Shrivastav *et al*[13] [2013]	5.27	2.44	68.5	23.2	34.11
Mondal *et al* [14] [2014]	5.7	2.5	73	22.7	30
Present study [2015]	4.1	2.31	64.4	17.9	27.7

Cases presented with severe anaemia, Reduced RBC Count, Reduced MCV, MCH, MCHC. Hemoglobin level is low compared to other studies, also MCV, MCH, MCHC are lower in present study because as patient who have received blood transfusion in last three months was excluded from study. Cases of Thalassemia intermedia are not included [as no case was found].

Table No. 20 Relevant Haematological Parameters in Group F [HbE-Beta Thalassemia]

Studies	Hb g/dl	RBC Million/mm3	MCV fl	MCH Pg	MCHC g/dl
Rao S *et al* [7] [2008]	6.2	3.2	63.9	16.3	30.1
Mondal *et al* [14] [2014]	7.7	3.5	65.2	18.7	29.4
Present study [2015]	3.4	2.12	59	16	27.1

Case in present study presented more severely [Low Hb] than other studies, as HbA0% was less in present study. Presentation of case depends to some extend on HbA0%[23]

Table No. 21 Relevant Haematological Parameters in Group [Hb Lepore Trait]

Studies	Hb g/dl	RBC Million/mm3	MCV fl	MCH Pg	MCHC g/dl
Rao S *et al*[7] [2008]	9.9	4.32	72.2	22.9	31.7
Mondal *et al*[14] [2014]	10.3	3.7	70.4	23.6	33.2
Present study [2015]	10.1	5.65	70.3	21.4	30.3

Haematological parameters of present study are consistent with Rao S *et al* study.

CONCLUSION

In the present study entitled "Study of Clinico-Hematological Profile of Hemoglobinopathies At Tertiary Care Centre", total 150 cases of clinically suspected hemoglobin disorders were studied by HPLC in the department of Pathology of Tertiary care hospital from October 2013 to October 2015.
HPLC was considered as the standard method.

1. Sickle cell trait was the most common Hemoglobinopathy detected.
2. Occurrence of Hemoglobinopathies was highest in Buddhas, followed by Muslim and Banjaras. Most common affected Ethnic group was Buddha followed by Banjaras in Sickle cell Syndromes
3. Pallor was the most common symptom in present study [49.33 %], followed by jaundice [13.33%].
4. Vaso-occlusive crisis [Abdominal pain, joint pain] was more common in sickle /Beta thalassemia than sickle cell anaemia, while jaundice was more common in Sickle cell anaemia than sickle /Beta thalassemia.
5. Jaundice was found to be less common in beta-thalassemia major.
6. Splenomegaly was more common in Beta thalassemia major than sickle cell anemia.
7. HPLC was found to be less labour intensive, rapid and more reliable for quantification of Hemoglobin variants.
8. Family studies are required to confirm the diagnosis.

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