



# STUDY OF SICKLE CELL ANEMIA IN HIGH INCIDENCE TRIBAL AREA BELT IN MADHYA-PRADESH- SHAHDOL DISTRICT

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## ABSTRACT

**Background:** Sickle cell disease (SCD) disproportionately impacts Adivasi (tribal) communities in India. Current research has focused on epidemiological and biomedical aspects but there has been scarce research on social determinants and health systems aspects. High prevalence of sickle gene has been demonstrated in various tribal communities of Madhya Pradesh including, patel, panica, baiga, gond, panihar, nagarachi, and kol etc. The present study was conducted with an objective to document the prevalence of sickle cell disease in tribal belt of central India population in shahdol, umaria, anooppur, mandla, dindori, katni and some population of pendra [chattisgarh] district of Madhya Pradesh state.

**Aims and objectives-** The study is conducted as a retrospective and prospective survey GMC Shahdol from September 2021 to march 2023, in 600 patients of sickle cell anaemia. All tribal patients who attended the opd and ipd of the hospital in routine or emergency, were screened for sickle cell anaemia by sickling test (with freshly prepared sodium metabisulphite). Mainly patients admitted in medicine, surgery and gynaecology department.

**Methodology:** The study is conducted as a retrospective and prospective survey GMC Shahdol from 2021 to 2022, were screened for sickle cell anaemia by sickling test (with freshly prepared sodium metabisulphite). Those found positive for sickling test, were further analysed by HPLC (high-performance liquid chromatography) to confirm their status as either sickle cell trait or sickle cell disease. All tribal patients who attended the opd and ipd of the hospital in routine or emergency, were screened for sickle cell anaemia by sickling test (with freshly prepared sodium metabisulphite). Mainly patients admitted in medicine, surgery and gynaecology department.

**Results:** Out of 600 cases number of male cases were 150 (25%) and females were 450 (75%) cases. The gender distribution of male to female cases is 1:3. 88.7% cases were sickle cell trait, 9.02% cases were sickle cell disease, 1.68% cases were sickle beta thalassemia, 0.25% cases were B Thalassemia Minor and 0.33% cases were B Thalassemia Major. In the present study, among the 600 cases. In which baiga [33%], panica [20%], panihar [8%], nagarachi [8%], kol [7%], patel [5%] and remaining are another cast. Tribal population affected around 70% patients of sickle cell anemia.

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**Conclusion:** It was concluded that patel, panica, baiga, gond, panihar, nagarachi, and kolwere the most common community of sickle cell disorders in tribal peoples Shahdol district. Among the Sickling test positive cases, Sickle cell trait and sickle cell disease was the most common variant.

**Keywords:** Sickle cell disease, Tribal community, Shahdol.

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## Introduction

The inherited disorders of blood include hemoglobinopathies which are one of the major public health problems in India [1]. Sickle cell disease is the second most common hemoglobinopathy next to Thalassemia in India [2]. There is a high prevalence of Sickle cell disease in the socio-economically backward groups in India. It is highly prevalent among Scheduled Caste, Scheduled Tribe, and Other Backward Class (10%) [3]. The management cost of these patients is very high and resources with Government are limited. Hence, the prevention appears to be the only solution in present circumstances. With a comprehensive medical care and management approach, the health status and life expectancy of these patients can be improved considerably. The high-risk couple for these disorders should be identified at the time ante-natal care and each pregnancy should be monitored. The couple should be given appropriate counselling after prenatal diagnosis

High prevalence of sickle gene has been demonstrated in various tribal communities of Madhya Pradesh including patel, panica, baiga, gond, panihar, nagarachi, and kol etc. The present study was conducted with an objective to document the prevalence of sickle cell disease in tribal belt of central India population in shahdol, umaria, anooppur, mandla, dindori, katni and some population of pendra [chattisgarh] district of Madhya Pradesh state.

The present study was conducted with an objective to document the prevalence of

sickle cell disease in Shahdol district of Madhya-Pradesh state.

## Materials and Methods

The study is conducted as a retrospective and prospective survey GMC Shahdol from September 2021 to march 2022, in 600 patients of sickle cell anaemia. All tribal patients who attended the opd and ipd of the hospital in routine or emergency, were screened for sickle cell anaemia by sickling test (with freshly prepared sodium metabisulphite). Mainly patients admitted in medicine, surgery and gynaecology department.

The blood was collected under all aseptic precautions. 2 ml of blood was drawn from antecubital vein by clean venepuncture from each patient with a sterile plastic syringe and collected in an EDTA (anticoagulant) tube for determination of investigations like Sickling test, Reticulocyte count. Those found positive for sickling test, were further analysed by HPLC (high-performance liquid chromatography) to confirm their status as either sickle cell trait or sickle cell disease.

Details were also recorded including age, sex, clinical and laboratory parameters. Categorical variable was expressed in actual number and percentages. Continuous variable was presented as Mean  $\pm$ SD.

## Results

There were 600 samples which were tested positive with Sickling test (DTT test) during September 2021 to march 2023.

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**Table-1: Age and gender distribution of cases**

Age (in years)	Male	Female	Total
0-10	30(20)	40(8.88)	70(11.66)

11-20	30(20)	125(27.77)	155(25.84)
21-30	34(22.66)	193(42.88)	227(37.83)
31-40	40(26.66)	50(11.11)	90(15)
41-50	10(6.66)	25(5.55)	35(5.83)
51-60	4(2.66)	12(2.66)	16(2.66)
>60	2(0.2)	5(1.11)	7(1.15)
<b>Total</b>	<b>150(25)</b>	<b>450(75)</b>	<b>600 [100]</b>

-there are 25% males and 75% female patients. Most common affected age group in males are 31-40 years and in females 21-30 years. The mean age of the cases participated in the study were 24.9 ± 13.134 years.

**Table-2: Types of sickle cell disease**

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Types of disease	No.	%
Sickle cell trait	532	88.7
Sickle cell disease	54	9.02
Sickle beta Thalassemia	10	1.68
B Thalassemia Minor	2	0.33
B Thalassemia major	2	0.33
<b>Total</b>	<b>600</b>	<b>100</b>

-among the 600 cases, 532(88.7%) cases were sickle cell trait, 54 (9.02%) cases were sickle cell disease, 10 (1.68%) cases were sickle beta thalassemia, 2 (0.25%) cases were B Thalassemia Minor and 2 (0.33%) cases were B Thalassemia Major.

**Table-3: Caste wise distribution of cases**

Caste	No.	%
Baiga	198	33.47
Panica	60	10
Panihar	48	8
Nagarachi	48	8
Kol	42	7
Patel	30	5

As far as caste distribution is concerned, among the 600 cases in which baiga [33%], panica [20%], panihar [8%], nagarachi [8%], kol [7%], patel [5%] and remaining are another cast. Tribal population affected around 70% patients of sickle cell anemia.

**Table- 4; sign and symptoms**

Sign and symptoms	Cases	Percentage
Anemia	300	50
Hepato-splenomegally	30	5
Gall bladder stone	18	3
Renal stone	6	1
Priapism	2	0.33



Ulcers	10	2
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-Anemia is most common presentation in these patients found in 50% cases. Hepatosplenomegaly in 5% cases, gall bladder stone in 3%, renal stone in 1%, ulcers in 2% cases and 2 cases also present with priapism.

**TABLE- 5; Maternal complications in SCA**

OUTCOMES	NO	PERCENTAGES
Abortion	60	10
Preterm birth	120	20
Painful chest synd.	6	1
Maternal death	6	1
Lscs	240	40
Painful crisis	180	30
Severe anemia	300	50
Jaundice	90	15
Infection	150	25
Pre-eclampsia	90	15

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-Severe anaemia most common observed complication in 50% cases followed by LSCS in 40% cases. Painful crisis in 30% cases, infection in 25%, preterm birth in 20%, jaundice in 15%, pre-eclampsia in 15%, abortion in 10% cases and painful chest syndrome and maternal death observed in 1% cases.

**Table – 6; fetal outcomes in SCA**

Fetal outcomes	No.	Percentages
Prematurity	240	40
Low birth weight	256	42
IUGR	48	8
Still birth	12	2
Neonatal death	18	3
5 min APGAR ≤7	36	6

-Low birth weight is most common, found in 40% cases. Prematurity found in 40% cases, IUGR in 8%, still birth in 2%, neonatal death in 3% and 5 min APGAR ≤7 in 6% cases found.

### Discussion

The prevalence rate of sickle haemoglobin in Madhya Pradesh ranges from 10% to 33% among different castes and tribal groups. There is a need to map the prevalence of this gene at micro level i.e. its variations in the different tribal groups and within a tribal group spread over a large area. Gond and Bhil group of tribals constitute a large proportion.

Among Gonds, the prevalence of sickle haemoglobin varies from 10% to 25% where as in the Bhils, the prevalence rate varies from 15% to 33%. Earlier studies carried out by various workers show that in Madhya Pradesh, the Scheduled Caste and Backward Class communities of the tribal predominant areas also have sickle cell gene in almost similar proportion (Unpublished reports). However, in



some scheduled caste populations, its prevalence is even higher than the adjoining tribal population. Hence, the problems of sickle cell gene in Madhya Pradesh exist among scheduled tribes, scheduled castes and backward class communities.

27 districts fall under sickle cell belt. These districts (arranged in descending order of ST & SC population) are Jhabua, Barwani, Dindori, Mandla, Dhar, Shahdol, Umaria, Betul, Seoni, West Nimar, Chhindwara, Harda, East Nimar, Jabalpur, Ratlam, Dewas, Katni, Damoh, Hoshangabad, Sagar, Satna, Balaghat, Ujjain, Indore, Mandsaur, Neemah and Narsimhpur.

Out of total 26 districts of Gujarat; more than half are tribal districts. Gujarat is the 4th most schedule tribe populated state of India after Madhya Pradesh, Maharashtra and Orissa. The tribal community of Gujarat inhabitants in the geographically difficult terrains of the Eastern belt, extending from Ambaji in the North to Dang in the South Southern Gujarat includes districts of Dangs, Valsad, Navsari, Surat and Bharuch[8].

Gujarat state is expected to have 6,47,025 Sickle trait and 48,257 Sickle disease patients, belonging to Dhodia, Dubla, Kukna, Gamit, Chaudhary, Halpati, Varli, Kokni, Kathodi, Kolcha, Kotwadia etc[9]. Sickle cell disease in this tribal area poses difficulty in diagnosis and management, as the sign and symptoms of this disease overlap with other common diseases. Recurrent attacks of musculoskeletal pain, anemia, frequent respiratory infections, jaundice and splenomegaly are the typical features which should arouse suspicion of sickle cell disease. We acknowledge that a community-based study is ideal to know the true prevalence of the disease but because of ethical/social issues involved, it is difficult to conduct a community-based study involving invasive procedure (blood collection). However, this hospital-based survey is important for sensitization and can serve as a baseline for generating more data.

-there are 25% males and 75% female patients. Most common affected age group in

males are 31-40 years and in females 21-30 years. The mean age of the cases participated in the study were  $24.9 \pm 13.134$  years.

-among the 600 cases, 532(88.7%) cases were sickle cell trait, 54 (9.02%) cases were sickle cell disease, 10 (1.68%) cases were sickle beta thalassemia, 2 (0.25%) cases were B Thalassemia Minor and 2 (0.33%) cases were B Thalassemia Major.

-As far as caste distribution is concerned, among the 600 cases in which baiga [33%], panica [20%], panihar [8%], nagarachi [8%], kol [7%], patel [5%] and remaining are another cast. Tribal population affected around 70% patients of sickle cell anemia.

-Anemia is most common presentation in these patients found in 50% cases. Hepatosplenomegaly in 5% cases, gall bladder stone in 3%, renal stone in 1%, ulcers in 2% cases and 2 cases also present with priapism.

Among the sickling test positive, 88.7% cases were sickle test trait. The higher prevalence of the sickle cell trait may be a result of a higher frequency of consanguineous marriages within the relatively small community. Association for Health Welfare in the Nilgiris (ASHWINI), Tamil Nadu also reported prevalence of sickle cell trait in non-tribal Chetti community to be as high as 30%[12]. Studies by S. L. Kate indicated that the overall prevalence of sickle cell disorder in different tribal populations is 10% for carrier state and 0.5% for the sufferer[13]. Sahu T et al reported 16.55% prevalence of sickle cell disorder in below fifteen years children in tribal areas of Gajapati district of Orissa [14].

-Severe anaemia most common observed complication in 50% cases followed by LSCS in 40% cases. Painful crisis in 30% cases, infection in 25%, preterm birth in 20%, jaundice in 15%, pre-eclampsia in 15%, abortion in 10% cases and painful chest syndrome and maternal death observed in 1% cases.

These are the main tribal communities of south Gujrat region. Sahu T et al has reported

Ratia, Sabar, Beera, Mandaletctribe community in their study in south Orissa [14]. The prevalence amongst the different communities in the decreasing order of frequency was Rathod (71.4%), Vasava (21.4%), Chavda (3.6%) and Solanki (3.6%) [15]. Saxena in his study reported prevalence of sickle cell anemia among Vasava was 26.4%, chaudhari 21.4%, Gamit 20%, Panchal 13.85% and among Rathod was 8.3% [16]. These observations support the hypothesis that the sickle cell disorders are present in scheduled castes, tribals and few communities of other backward classes (OBC), and not found in so called higher castes; though the review of literature says it is present invariably in all castes [17].

-Low birth weight is most common, found in 40% cases. Prematurity found in 40% cases, IUGR in 8%, still birth in 2%, neonatal death in 3% and 5 min APGAR  $\leq 7$  in 6% cases found.

Majority cases were found positive for sickle cell trait disease belongs to Dhodia community (33.47%), while Dalal M has reported 18.4% prevalence of sickle cell trait in Dhodias of south Gujarat region [18]. The variation in proportion of people with different communities dependson their distribution. In Gujarat, the Dhodia, Dubla, Gamit, and Naikatribes have a high prevalence of HbS (13-31%)[19]. More recently very extensive population surveys have been done by the Indian Red Cross Society, Gujarat State Branch where 1,68,498 tribals from 22 districts were screened and the overall prevalence of sickle cell carriers was 11.37% [20]. Some tribal groups in south Gujarat like Chaudry, Gamit, Rohit, VasavaandKukanahave shown both a high prevalence of HbS (6.3 to 22.7%) as well as  $\beta$ -thalassaemia trait (6.3 to 13.6 %).[21] These tribal groups would have the likelihood of co-inheriting both these genes.

### Conclusion

With the advances in molecular genetics, it is possible to detect the defect at early stage (10 to 15 weeks) of pregnancy. The management

cost of these patients is very high and resources with Government are limited. Hence, the prevention appears to be the only solution in present circumstances. With a comprehensive medical care and management approach, the health status and life expectancy of these patients can be improved considerably. The high-risk couple for these disorders should be identified at the time ante-natal care and each pregnancy should be monitored. The couple should be given appropriate counselling after prenatal diagnosis. The facilities and technical know-how for diagnosis of the disorder and its Prevention and management: clinical management should be generated at PHC/ district hospital level depending upon the disease load. There should be at least two to three genetic counselling centres in the state for prenatal diagnosis and counselling. The State Government should have comprehensive plan for prevention and management of the sickle cell disease.

It was concluded that patel, panica, baiga, gond, panihar, nagarachi, and kol were the most common community of sickle cell disorders in tribal peoples of Valsad Shahdol district. Among the Sickling test positive cases, Sickle cell trait and sickle cell disease was the most common variant.

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