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Original Article

SICKLE CELL DISEASE IN JHABUA AND KHARGONE DISTRICT: UNVEILING PREVALENCE AND SEVERITY

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ABSTRACT

Objective: To assess the prevalence among Sickle cell disease (SCD) affected individuals emphasizing the neglected health challenges in various tribes.

Methods: Cross-sectional, observational study was conducted during the district residency program for 9 mo. The data has been collected from the record room of patients diagnosed with Sickle cell Anemia. Statistical analysis was done using Microsoft Excel.

Results: A total of 295 patients' data revealed demographic skew toward Jhabua (50%), with Sickle cell anemia diagnosed at the mean age of 23±3.9. Most patients (72.3%) were Hindu, with Bhil and Bhilaya tribes having higher frequencies. Symptoms varied; 94% had Sickle cell trait, 16.3% had sickle cell disease, and 60% experienced painful crises. Treatment included prophylactic care for all, 37.57% required blood transfusions and 29.7% were on hydroxyurea.

Conclusion: The study underscores the significant SCD burden and the need for heightened awareness and targeted interventions in socio-economically disadvantaged tribal regions to mitigate the impact of SCD.

Keywords: Sickle cell disease, Prevalence, Tribal population, Treatment pattern, Disease burden

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INTRODUCTION

Sickle cell disease (SCD) is a significant global health challenge despite receiving less attention compared to infectious diseases. SCD is the most common genetic disease in the United States, affecting 1 in 500 African Americans, with approximately 300,000 infants born with sickle cell anemia annually [1]. The disease is characterized by multisystemic complications, and its severity can vary due to genetic mutations and modifier genes [2]. Despite substantial efforts directed toward combating infectious diseases such as malaria, tuberculosis, and HIV, interventions aimed at addressing birth defects, particularly genetic disorders, have received less attention. It is estimated that over 7 million babies are born annually with congenital abnormalities or genetic diseases, with hemoglobinopathy and glucose-6-phosphate dehydrogenase deficiency constituting 25% of these cases [3]. SCD, an autosomal recessive genetic condition resulting from mutant hemoglobin genes, manifests in sickle cell trait or symptomatic sickle cell anemia (SCA) disease [4]. The disease's impact is characterized by sickle-shaped red blood cells, leading to anemia, sickle cell crisis, and reduced tissue perfusion. Although a century has passed since its discovery, affordable cures for SCD remain elusive, emphasizing the need for preventive measures. In India, where the sickle cell gene prevalence varies among different ethnic groups, effective management strategies, especially in tribal populations, are critical.

According to the Census of India 2011, the prevalence of tribal population in India is 8.6 % of the total population. The states of Madhya Pradesh, Maharashtra, Odisha, Gujarat, Rajasthan, Jharkhand, Chhattisgarh, Andhra Pradesh, West Bengal, and Karnataka account for around 83 percent of the total scheduled tribe population in the country and the majority of these tribal groups live in rural areas [5]. A study conducted by Utkarsha S. *et al.*, mentioned that Hemoglobin S is majorly widespread in the tribal populations of Southern, Central, and Western states, with prevalence rates reaching as high as 48% in certain communities [6]. The presence of sickle hemoglobin in blood among the Indian population was first described by Lehman and Cutbush in 1952 in the tribal populations of Nilgiri Hills, South India. In the same year, Dunlop and Mazumder also reported the presence of sickle hemoglobin in the tea garden workers of Upper Assam, who were migrant labourers from tribal groups in Bihar and Odisha [7].

Madhya Pradesh has the greatest burden of this disease, with an estimated number of 9,61,492 sickle heterozygotes and 67,861 sickle homozygotes. More than half of the districts in Madhya Pradesh fall under the sickle cell belt, and the prevalence of HbS varies from 10-33%. In the previous study done by Colah RB *et al.* it was estimated that 13,432 pregnancies would be at risk of having a child with sickle cell disease in this State, and the expected annual births of sickle homozygotes would be 3358 for the year 2015. The largest tribal groups in Central India consist of *Gonds* and *Bhils* [8]. This study aims to assess the current prevalence and severity of carriers and sufferers of SCD during the District Residency Program by NMC in the Jhabua and Khargone districts of Madhya Pradesh. Given the influential role of socio-economic factors in the progression of the disease, raising awareness among populations in impoverished and educationally deprived areas, particularly within remote tribal regions, is crucial to achieving favourable outcomes in the future.

MATERIALS AND METHODS

A cross-sectional prospective study was carried out in the Jhabua and Khargone Districts of Madhya Pradesh by the postgraduate residents of the Pharmacology Department of MGM Medical College, Indore MP, under the National Medical Commission's DRP Programme. The study spanned a period of nine months from April 2023 to December 2023 and it was conducted after obtaining due permission from the Chief Medical and Health Officer (CMHO) of district hospitals Jhabua and Khargone. Convenient sampling was employed in the aforementioned districts, and the recorded data of sickle cell disease patients of all age groups registered in the district hospitals were included in the study, resulting in a total of 295 participants. Data of the patients presenting with other congenital diseases were not included in the study. The study involved the

acquisition of socio-demographic data encompassing variables such as sex, religion, health status, and information regarding the treatment of their illnesses, history of blood transfusions, and complications. The study participant's weight and height were measured using a stadiometer and weighing scale, respectively, and their body mass index (BMI) was calculated utilizing the standard formula. The confidentiality of patients was ensured throughout the study. The data was recorded in MS Office Excel 2011 and analyzed using frequencies, percentages, and proportions.

RESULTS

Demographic distribution

More than 50% of the patients were from Jhabua as compared to Khargone. The mean age of diagnosis of SCA among study patients was 23±3.9 y from the young adult age group. Female were 65.9%. On calculating their BMI, 182 (61.5%) patients were found underweight, while only 33.5% of them were within the normal range.

Table 1: Demographic distribution among sickle cell disease

1.	Age	Frequency (N=295)
	0-20 y	121 (41.3%)
	21-40 y	155 (52.3%)
	≤40 y	18 (6.2%)
2.	Gender	
	Male	100 (34.1%)
	Female	195 (65.9%)
3.	BMI	
	Underweight	182 (61.5%)
	Normal	99(33.5%)
	Overweight	12 (4%)
	Obese	2 (1%)

Community-wise distribution

The majority of patients, numbering 213 (72.3%), belong to the Hindu religion among the various ethnic tribal groups; the highest frequency of the disease was observed in Bhil (26.1%), Bhilaya (21.5%), and Pateliya (14.7%), while the lowest frequency was found in Gond (10%). Muslims and Buddhists account for 13.8% each. A higher number of patients were obtained from the Scheduled Caste (63.1%) followed by Scheduled Tribes (28.8%) and Other Backward Classes (6%), while 2.1% belonged to the open category.

Occurrence of disease and its complication

In our study of 295 cases, 94% exhibited Sickle cell trait (heterozygous), while 16.3% had sickle cell disease (homozygous). All patients experienced multiple responses, including hemolytic anemia. Significantly, 60% endured painful crises, 59% had recurrent infections, 46% developed jaundice, and others faced complications like gallbladder stones and renal complications.

Treatment outcomes

The treatment review revealed that all 295 patients received regular prophylactic care, which included Folic acid and Vitamin C supplements. Among those patients who required regular blood transfusions, 37.57% received them. Additionally, 29.7% of patients were undergoing Hydroxy Urea treatment. Interestingly, 13% of patients showed a significant increase in hemoglobin gram per cent (Hb gm%) without needing blood transfusions or Hydroxyurea treatment. On the other hand, 8.7% of patients did not respond to the treatment. Moreover, 11.03% of patients had to be hospitalized for various morbidities.

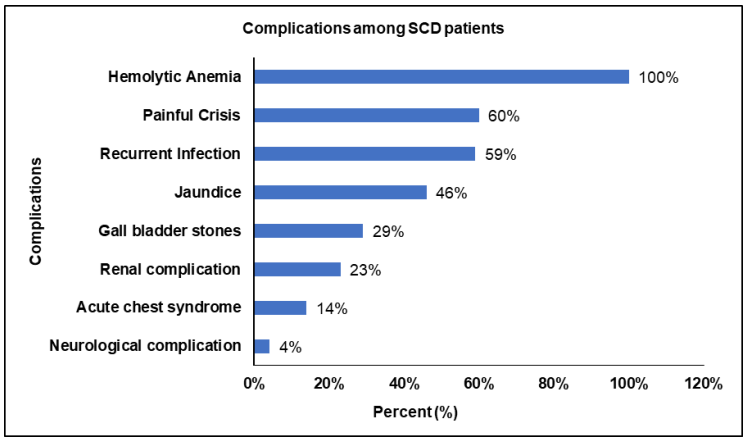

Table 1: Complications like gallbladder stones and renal complications

DISCUSSION

The results of the study show diagnosis of sickle cell anemia (SCA) was a higher prevalence in the young-adult age group. In a previous study, the mean age of SCD patients was 26.34±8.19 y [9]. Body mass index (BMI) showed that 62% of the patients were underweight, while the remaining patients had a normal BMI. Contrasting results were found in the study conducted by Jackson *et al.*, which revealed raised BMI [10].

The majority of patients with sickle cell anemia in the study belonged to the Hindu religion, with the highest frequency of the disease observed in the Bhil, Bhilaya, and Pateliya ethnic tribal groups and the lowest frequency in the Gond group. Among the different caste categories, the highest number of patients was from the Scheduled Caste, followed by Scheduled Tribes and Other Backward Classes, while a smaller percentage belonged to the open category. This ratio is comparable with other studies and varies in different districts and states [11].

In our study of 295 cases, we found a high prevalence of sickle cell trait among 94% of individuals (heterozygosity). This surpasses a study by Piel *et al.* (2013), in which the global incidence of the sickle trait birth was approximately three percent [12]. However, this substantial variation may stem from our study population's geographic and genetic characteristics that may predispose to this genetic abnormality. Of the total patients,16.3% exhibited sickle cell disease (homozygous), a percentage consistent with the global prevalence of the disease. The most common complications observed were painful crises (60%), recurrent infections (59%), jaundice (46%), gallbladder stones, and renal complications. These findings align with another study by Ballas *et al.* (2010), which also reported painful crises and infections as common complications [13]. Regarding treatment, all patients received regular prophylactic care, including Folic acid and Vitamin C supplements, similar to a study by Vichinsky *et al.* which advocated for regular prophylactic care in managing sickle cell disease [14]. Moreover, 37.57% of our patients required regular blood transfusions, and 29.7% were undergoing Hydroxy Urea treatment. In a study done by Praharsh KMR *et al.*, it was mentioned that long-term use of hydroxyurea has multiple hematological and dermatological adverse effects [15]. Interestingly, 13% of patients demonstrated a significant increase in hemoglobin gram per cent (Hb gm%) without needing blood transfusions or Hydroxyurea treatment, suggesting that some patients may have a milder form of the disease or may respond better to the prophylactic care.

However, it's worth observing that 8.7% of patients did not respond to the treatment, and 11.03% had to be hospitalized for various morbidities, underlining the severity and unpredictability of the disease. In comparison to a study by Ware *et al.* (2017), which reported a non-response rate of approximately 10%, our study findings are in line with previous research done [16].

LIMITATIONS

The study includes a nine-month duration, aligned with the District Residency Program rotations of three batches. As a result, the findings may not be easily generalized due to the specific timeframe and cohort involved.

CONCLUSION

Our study reveals a higher incidence of sickle cell disorders in the Jhabua region compared to Khargone, with a significant number of patients being young-adult and underweight. Predominantly, we observed the prevalence of these disorders among individuals of the Hindu faith, with ethnic groups such as the Bhil, Bhilaya, and Pateliya demonstrating higher disease rates. A wide array of health issues was observed among the patients, with hemolytic anemia and painful crises being particularly common. All patients were provided with prophylactic care, including supplements of Folic acid and Vitamin C. A noteworthy segment of the patient population also underwent blood transfusions or were treated with Hydroxyurea. While some patients responded positively to the treatment protocols, others faced challenges, underscoring the complex nature of these disorders. This study emphasizes the importance of future research endeavours aimed at deciphering the factors that influence non-response in certain patients. Additionally, there is a pressing need to develop personalized treatment strategies that cater to individual patient needs, potentially improving disease management outcomes. There should be an introduction of the program to screen for sickle cell trait and also to provide genetic counselling before marriage so as to reduce the prevalence of sickle disease in these niche populations.

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Nil

ABBREVIATION

SC: Schedule Caste; ST: Schedule Tribe; SCD: Sickle Cell Disease; DRP: District Residency Programme.

AUTHORS CONTRIBUTIONS

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Data collection: Dr Avina Kharat, Dr. Ruchi Kumari, Dr. Paroma Sinha, Dr. N. Vignan, Dr. Akash Mishra, Dr. Siddharth Ojha.

Writing-original draft preparation-Dr. Avina Kharat.

Final review-Dr. Anjali Kushwah, Dr. Ruchi Kumari.

CONFLICT OF INTERESTS

Declared none

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