Research Article

A study to assess the socio-epidemiological profile of sickle cell disease among affected tribal population

Dilip Dhaku Kadam[1], Aritra Kumar Bose[2], Prashant Laxmanrao Chandekar[3]

[1]Department of Community Medicine, Seth Gordhandas Sunderdas Medical College and King Edward Memorial Hospital, Mumbai, Maharashtra, India, [2]Department of Community Medicine, Grant Government Medical College and Sir Jamshedjee Jeejeebhoy Group of Hospitals, Mumbai, Maharashtra, India, [3]Department of Community Medicine, Government Medical College, Chandrapur, Maharashtra, India

Correspondence to: Prashant Laxmanrao Chandekar, E-mail: dr.prashant.chandekar.100@gmail.com

Received: August 25, 2019; **Accepted:** September 21, 2019

ABSTRACT

Background: Sickle cell disease (SCD) is more than a century old disease, but we still do not have an affordable cure for it. Although studies show that the prevalence of the disease is high in tribal areas, awareness and mainstreaming of management of SCD in the Primary Health Cares are still not formalized in the system. Our study aims to provide practical insights for imparting quality services and ensure that the provisions reach at the doors of patients. **Objectives:** The objectives of the study were to study the epidemiological profile and clinical patterns of SCD among affected tribal population. **Materials and Methods:** The study was conducted at a tribal area named Sakwar. By applying suitable formula, the sample size was calculated to be 197. After obtaining informed consent, one to one interview method was used for data collection regarding health status, previous diagnosis, complication, etc. Body mass index of the patients was calculated using standard formula. SPSS version 22 was used for statistical analysis. **Results:** Majority of the subjects were females (62.44%) belonging to Hindu religion 145 (73.60%) and scheduled tribes (67.01%). The study subjects were predominantly students belonging to lower socioeconomic status. About 67% of patients were having sickle cell trait while 33% had SCD. Among complications 50% had painful crisis, 36% had gallbladder stone, 15.58% had jaundice, and 5.08% study subjects had avascular necrosis. About 18.7% of study subjects had to receive blood transfusion for their disease. About 18.27% of study subjects were underweight. Majority of the study subjects were taking treatment from private practitioner while government hospitals were preferred for inpatient care and blood transfusion. We found SCD occurrence was significantly associated with female sex and consanguineous marriage. Receiving treatment from private practitioners was significantly associated with occurrence of complication among the study subjects. **Conclusion:** The study found that the disease and its complication were significantly more among female sex. Occurrence of complication was more common among people taking treatment from general practitioner.

KEY WORDS: Anemia; Genetic Diseases; Health Services; Indigenous; Inborn; Rural Health; Sickle Cell

Access this article online	
Website: http://www.ijmsph.com	Quick Response code
DOI: 10.5455/ijmsph.2019.0926921092019	

INTRODUCTION

Sickle cell disease (SCD) holds the distinction of being the first inherited disease identified at the molecular level. While considerable efforts are currently being invested into reducing the global burden of infectious diseases, particularly malaria, tuberculosis, and HIV,[1] interventions to decrease

birth defects have largely been neglected.[2] It has recently been estimated that more than 7 million babies are born each year with either a congenital abnormality or a genetic disease.[2] Five disorders constitute approximately 25% of these births, two of which are monogenic diseases, namely, hemoglobinopathy and glucose-6-phosphate dehydrogenase deficiency.[2]

SCD is a hemoglobinopathy which is an autosomal recessive genetic condition caused due to inheritance of mutant hemoglobin (Hb) genes. The disease occurs in two forms, a milder form called sickle cell trait (symptomless) and a symptomatic form called the sickle cell anemia (SCA)/ disease. Due to abnormal genes, the red blood cells (RBCs) do not contain enough Hb or contain abnormal Hb called Hbs. As a result the RBCs which are normally circular in shape, become sickle-shaped when exposed to low oxygen tension. Normal RBC's smoothly pass through blood vessels without any obstruction, but sickle cells cannot pass easily and rupture resulting in moderate-to-severe anemia in such patients. Due to continued rupture and polymerization of the defective RBC's, blockage of the small blood vessels occur which leads to diminished tissue perfusion causing severe pain and swelling. This is termed as sickle cell crisis.[3]

Even though a century has passed after the discovery of the disease, we still do not have an affordable cure for it. The only treatment for the disease is bone marrow replacement which is costly although having a poor success rate. Till date, prevention of complications is the only cost-effective modality which is available to us. Unfortunately, many tropical countries do not have the necessary resources required to provide the complex care required for SCD patients, and resulting outcomes are typically poor. Patients with SCD generally have a median survival of <5 years under conditions of limited medical resources.[4] While in high-income settings the current life expectancy for patients with SCA is estimated to be between 45 and 55 years of age, in low- and middle-income countries it is thought that most children die before reaching adulthood, with more than 500 children with SCD dying every day because of poor access to appropriate treatment.[2,5]

In India, many population groups have been screened and the sickle cell gene has been shown to be prevalent among three socioeconomically disadvantaged ethnic groups, the scheduled tribes (ST), scheduled castes, and other backward classes (OBC).[6,7] Although the prevalence of sickle cell varies from 1 to 40% among various tribal groups in India,[7] the mainstreaming of management of SCD in the Primary Health Cares is not formalized in the system. Further, the socio-cultural practices and realities compound the challenges especially in the view of tribal population, which is known to be scattered in a given geographic area. Under these circumstances, in-depth study of epidemiological factors may provide practical insights for imparting quality services and ensure that the provisions reach the doors of patients. The currents study would help us in early detection of cases and plan the intervention programs effectively, so as to reduce the burden of disease and improve quality of life of the SCD patients.

We aim to study the epidemiological profile and clinical patterns of SCD among the tribal population in this article.

MATERIALS AND METHODS

The study was conducted at a tribal area named Sakwar, which is the rural field practice area of Seth Gordhandas Sunderdas Medical College, Mumbai. Approval for conduction of study was obtained from the Institutional Ethics Committee. By applying suitable formula, the sample size was calculated for the most common morbidity that, i.e., anemia in general population (50% prevalence) as it is most common morbidity in SCD. The final sample size came to 197. A NGO working on SCD, in the study area was contacted, and their help was sought in getting the names and address of the SCD patients in the area. Random number table was used to recruit the patients in the study. The study was conducted over a period of 6 months that is from June 2018 to November 2018.

A semi-structured, pre-validated questionnaire was prepared in accordance with the objectives of the study. All selected patients in the study area were visited individually. Informal discussion was done with them to build rapport. The purpose of the study was explained using an informed consent document, and such consent was obtained from each of the study subjects in Hindi. A time schedule for the interview was prepared in consultation with the family members giving due consideration to the feasibility of their availability at home. One to one interview method was used to interview patients after taking consent, and the data were collected with the help of a pre-validated questionnaire. Respondents were asked about their health status, previous diagnosis, and any past consequences of SCA. The information about treatment of illnesses, place of treatment, and blood transfusion history was obtained from their medical records. Weight and height of the patients were measured using stadiometer and weighing scale, respectively. Body mass index (BMI) was calculated using standard formula.

Confidentiality of the study subjects was maintained. Statistical analysis of the data was done using SPSS version 22. Association between different variables was analyzed using Chi-square test wherever applicable.

RESULTS

The study subjects were predominantly from young age group (n = 22.17 years). Among them, 74 (37.56%) were males and 123 (62.44%) were females. Majority 145 (73.60%)

subjects belonged to Hindu religion while 52 (26.40%) were Buddhist. Majority of the patients were ST (67.01%) followed by the schedule cast (SC) (26.90%) and OBC (5.08%) while 2.02% belonged to open category. About 197 (99.44%) study subjects were literate while only 7 (3.55%) were found to be illiterate. On enumerating their occupations 118 (59.01%) of the subjects were students while 55 (27.9%) were laborers, other professions in which the study subjects were involved are business, 11 (5.6%), independent profession 5 (2.5%), and 1 (0.5%) were in service. Majority 149 (74.16%) of the subjects belonged to lower SE class according to the Pareek's socioeconomic classification. It was seen that majority, i.e., 103 (51.27%) subjects were married, among them 27 (13.71%) subjects had consanguineous marriage, i.e., marriage within first-order relatives.

On looking into the clinical profile of patients, we found that 134 (67%) patients were having sickle cell trait, i.e., heterozygous state while 66 (33%) had SCD, i.e., homozygous state. History revealed the sickle cell status of parents of patients [Table 1]. On calculating their BMI, 37 (18.27%) study subjects were found to be underweight while the rest were within normal range.

Table 1: Distribution of SCD patients according to disease among their parents

Father	No. of cases	Percentage
Trait	33	16.75
Disease	6	3.05
Not know	158	80.20
Mother		
Trait	17	8.63
Disease	11	5.58
Not know	169	85.79

SCD: Sickle cell disease

It was observed that all of the subjects had history of hemolytic anemia and 50% had painful crisis [Figure 1]. Scrutiny of case papers of study subjects revealed that 36% developed gallbladder stone, 15.58% had developed jaundice, and avascular necrosis (AVN) was found in 5.08% study subjects. It was seen that 18.7% study subjects had to receive blood transfusion for their disease and three subjects had to receive it thrice a year while one received 5 times a year.

Treatment profile of the patients revealed that most of the patients116 (58.88%) were consulting a general practitioner (GPs), 34 (17.14%) received treatment in primary health center while 50 (25.88%) were taking treatment from district hospital/tertiary health care. Out of 90 subjects who required hospital admission for the management of morbidities, 23.9% availed treatment from government hospitals, 15.2% from private hospital while other was admitted in both hospitals. Among the patients who needed regular blood transfusion, 67.57% received it from government setups while the rest visited private facilities to get the same. Loans were availed by 11% subjects for bearing the treatment expenses.

The study findings indicate that the occurrence of SCD was significantly associated with sex ($X^2 = 2.46$, $P = 0.001$) while occurrence of complications had no significant relationship with the sex of the study subjects. The occurrence of SCD was significantly associated with finding of consanguineous marriage among study subjects ($X^2 = 2.78$, $P = 0.04$). Place of treatment was significantly associated with occurrence of complication among the study subjects ($X^2 = 3.27$, $P = 0.04$) [Table 2].

DISCUSSION

Majority of the subjects were females (62.44%) belonging to Hindu religion 145 (73.60%) and ST (67.01%). The study

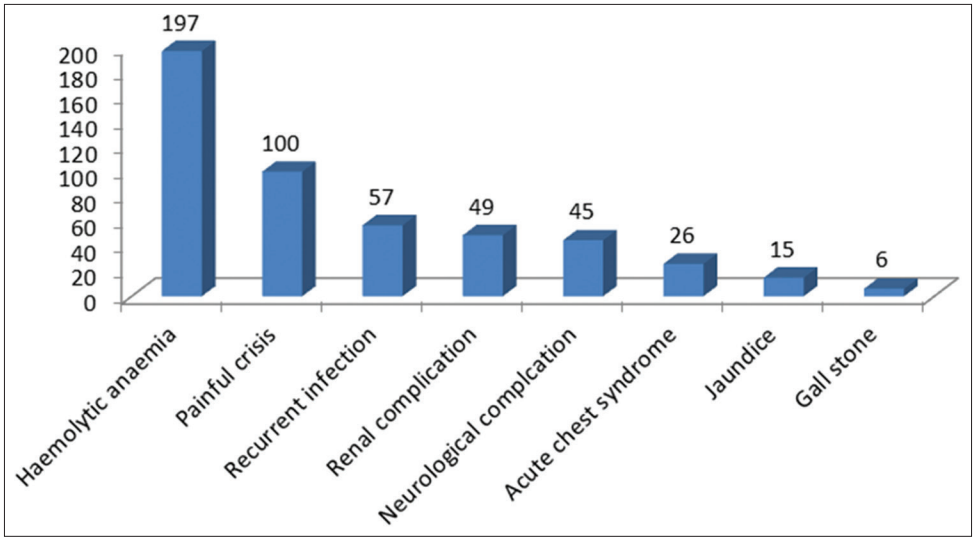

Figure 1: Complications among sickle cell disease patients (multiple responses given)

Table 2: Association between SCD and epidemiological factors

Variables	N (%)	χ^2, DF	*P* value*
Sickle cell anaemia			
Male	14 (21.5%)	9.23,1	0.001
Female	50 (78.46%)		
Complication of sickle cell anaemia			
Male	20 (43.09%)	0.26,1	0.52
Female	27 (57.37%)		
Consanguineous marriage In SCD			
Yes	28 (77.78%)	2.78,1	0.04
No	8 (22.22%)		
Complication with treatment from GP			
Yes	83 (65.87%)	3.27,1	0.04
No	33 (26.19%)		

**P*<0.05=Significant

subjects were predominantly students belonging to lower socioeconomic status. About 67% of patients were having sickle cell trait, while 33% had SCD. About 18.27% of study subjects were underweight. The subjects experienced complications like painful crisis, gall bladder stone, jaundice and AVN. Few of the study subjects had to receive frequent blood transfusion. Majority of the study subjects were taking treatment from private practitioner while government hospitals were preferred for inpatient care. We found SCD occurrence was significantly associated with sex and consanguineous marriage. Place of receiving treatment was significantly associated with occurrence of complication among the study subjects.

The study population consisted of predominantly females belonging to the economically productive age group, although there was no significant relationship between occurrence of the disease and sex of the subjects being female. In an article by Garg *et al*.,[8] it was reported that the prevalence of SCD was more among females than in males. We found patients were predominantly from SC and ST caste belonging to the Buddhist community. Similar findings were reported by Gunjal *et al*.[9] and Deshmukh *et al*.[8] who reported a higher prevalence of SCD among SC, ST caste, and tribal people. The patients in our study were from low socioeconomic status, and some had to avail loans for getting treatment for SCD. Our study findings indicate that although frequencies of consanguineous marriages were less among the study population, consanguineous marriages were significantly associated with occurrence of SCD. Daak *et al*.[10] reported very high frequency of consanguineous marriages (67.5%) in their study on sickle cell patients. Furthermore, Mouzan *et al*.[11] reported in their study that sickle cell was the second most common genetic disorder caused due to consanguinity. This can be explained by the fact that tribal people have unicentric geographical origin and endogamy among them leads to high frequency of occurrence of mutated gene in their genotype. We found more patients of sickle cell trait than SCD in our study which is similar to the finding reported by Karadağ *et al*.[12] and Mandot *et al*.[13] Anemia was present in all the patients and about a quarter of patients were underweight. Other symptoms reported by the patients were painful crisis, recurrent infection, renal complications, etc. Mukherjee *et al*. in their study reported similar finding of moderately high prevalence of malnutrition among tribal population. Furthermore, Charuhas *et al*.[14] reported very high prevalence of anemia among the tribal community in their study. Similarly, Khapre *et al*.[15] found in the study that the prevalence of anemia among rural women was 86%. These points toward anemia due to sickling may be getting aggravated due to inadequate nutrition intake which may be due to unawareness or financial reasons, although we did not find any significant relationship between the same. Similar to our study Elmoneim *et al*.[16] found that acute painful crisis, acute chest syndrome, and recurrent infection were the most common symptoms in SCD patients in their study. We found that the most common complication reported by the patients was gallstones followed by jaundice and AVN. Also, the occurrence of complication was significantly more among females. Esezobor *et al*.[17] reported complication of stroke and AVN in their study. The difference in complication profile in the present study may be due to difference in pattern of food intake and environmental influence among the study population. Patients in our study preferred GPs as compared to government hospitals in receiving medical advice although government hospitals were preferred for inpatient care and blood transfusions. A statistically significant relationship was established between source of treatment and occurrence of complication which indicates that the odds of developing complication was more when treatment was taken from GPs as compared to government hospitals.

The current study is an attempt to capture a snapshot of SCD patients, which points toward the unawareness and financial burden which the disease puts on patients. There may have been deliberate under-reporting by the respondents as the data were self-reported. Furthermore, further cohort studies can be undertaken to establish causal relationship between risk factors and the morbidities enumerated in the study.

CONCLUSION

SCD is X-linked recessive disease, which is genetically passed from generation to generation. The disease with its treatment being blood transfusion, bone marrow transplant, and costly drugs is beyond the reach of general population. Its complications lead to frequent hospital admission leading to unwarranted out of pocket expenditure by family. Due to this financial burden, people are pushed below poverty line. Furthermore, loss of man hours results in daily wage loss adding to the financial burden. Hence, prevention is the most important aspect in combating the problem of SCA. Thus,

SCD awareness and detection campaigns should be intensified in the vulnerable districts and talukas so that the patients have information regarding availability of appropriate treatment. The government health facilities must be upgraded to meet clinical management needed for the disease. Continuing medical education must be conducted regularly to help the GPs in updating their knowledge so that they could provide appropriate management so as to prevent complication and increase awareness regarding the disease. Furthermore, the National Social Service scheme must be implemented in rural and tribal area colleges for creating awareness about SCD and promote blood donation campaigns so that safe blood is available to the patient during crisis. Economic and travel concession schemes for sickle cell patients and their accompanying relative should be given wide publicity among tribal populations so as to decrease economic burden on the patients. Thus, a comprehensive approach comprising all level of prevention is needed to tackle the problem of SCA at the community level.

REFERENCES

1. Katz I, Komatsu R, Low-Beer D, Atun R. Scaling up towards international targets for AIDS, tuberculosis, and malaria: Contribution of global fund-supported programs in 2011-2015. PLoS One 2011;6:e17166.
2. Piel FB, Hay SI, Gupta S, Weatherall DJ, Williams TN. Global burden of sickle cell anaemia in children under five, 2010-2050: Modelling based on demographics, excess mortality, and interventions. PLoS Med 2013;10:e1001484.
3. Lervolino LG, Baldin PE, Picado SM, Calil KB, Viel AA, Campos LA. Prevalence of sickle cell disease and sickle cell trait in national neonatal screening studies. Rev Bras Hematol Hemoter 2011;33:49-54.
4. Kunz JB, Cario H, Grosse R, Jarisch A, Lobitz S, Kulozik AE. The epidemiology of sickle cell disease in germany following recent large-scale immigration. Pediatr Blood Cancer 2017;64:e26550.
5. Weatherall D. The inherited disorders of haemoglobin: An increasingly neglected global health burden. Indian J Med Res 2011;134:493-7.
6. Rao VR. Genetics and epidemiology of sickle cell anemia in India. Indian J Med Sci 1988;42:218-22.
7. Kaur M, Das GP, Verma IC. Sickle cell trait and disease among tribal communities in Orissa, Madhya Pradesh and Kerala. Indian J Med Res 1997;105:111-6.
8. Garg B, Garg N, Prajapati N, Bharambe M, Deshmukh P. Prevalence of sickle cell disorders in rural Wardha. Indian J Community Med 2006;31:26.
9. Sandeep SG, Uday WN, Arun YH. Prevalence of sickle cell disorder and anaemia in tribal school students from central India. Int J Collab Res Intern Med Public Health 2012;4:1321-9.
10. Daak AA, Elsamani E, Ali EH, Mohamed FA, Abdel-Rahman ME, Elderdery AY, *et al.* Sickle cell disease in Western Sudan: Genetic epidemiology and predictors of knowledge attitude and practices. Trop Med Int Health 2016;21:642-53.
11. El Mouzan MI, Al Salloum AA, Al Herbish AS, Qurachi MM, Al Omar AA. Consanguinity and major genetic disorders in Saudi children: A community-based cross-sectional study. Ann Saudi Med 2008;28:169-73.
12. Karadağ G, Güngörmüş Z, Olçar Z. Experiences and problems encountered by families of children with sickle cell anemia. J Caring Sci 2018;7:125-9.
13. Mandot S, Khurana VL, Sonesh JK. Sickle cell anemia in garasia tribals of Rajasthan. Indian Pediatr 2009;46:239-40.
14. Charuhas VA, Neelam DS, Sanjay SK, Sanjay BA, Mohan BK, Sanjeev MC, *et al*. Do gender differences influence the prevalence of sickle cell disorder and related morbidities among school children in rural central India? Int J Collab Res Intern Med Public Health 2009;5:14-18. Available from: http://www.internalmedicine.imedpub.com/do-gender-differences-influence-the-prevalence-of-sicklecell-disorder-and-related-morbidities-among-schoolchildren-in-rural-central-india.php?aid=6300. [Last accessed on 2019 Jul 18].
15. Khapre M, Meshram R, Mudey A. Study of multiple casual factors associated with varying degree of anaemia among rural pregnant women. Int J Med Sci Public Health 2013;2:268.
16. Abd Elmoneim AA, Al Hawsawi ZM, Mahmoud BZ, Bukhari AA, Almulla AA, Sonbol AM, *et al.* Causes of hospitalization in sickle cell diseased children in Western Region of Saudi Arabia. A single center study. Saudi Med J 2019;40:401-4.
17. Esezobor CI, Akintan P, Akinsulie A, Temiye E, Adeyemo T. Wasting and stunting are still prevalent in children with sickle cell anaemia in Lagos, Nigeria. Ital J Pediatr 2016;42:45.

How to cite this article: Kadam DD, Bose AK, Chandekar PL. A study to assess the socio-epidemiological profile of sickle cell disease among affected tribal population. Int J Med Sci Public Health 2019;8(12):1029-1033.

Source of Support: Nil, **Conflict of Interest:** None declared.