

Situational Analysis of Sick Cell Disease in Maharashtra, IndiaSunmeet Matkar¹, Yogesh Nawar²¹Senior Executive, Medical Communication, Metropolis Healthcare Ltd²Product Manager, Metropolis Healthcare Ltd, Mumbai, India.**Correspondence Author:** Sunmeet Matkar, Senior Executive, Medical Communications, Metropolis Healthcare, India**E-Mail Id:** matkar.sunmeet@gmail.com**Conflicts of Interest:** Nil**Abstract**

Sickle cell disease (SCD) is the foremost public health apprehension, not only in Maharashtra but also on global stage. Maharashtra, a western state of India, has 96.87 lakh tribal populations and is expected to have at least 11,00,000 sickle cell trait and 90,000 SCD patients. An objective of this review is to determine the prevalence of SCD in various communities in Maharashtra and various screening methods employed.

Materials and Methods

An in-depth literature review was undertaken with the help of available search engines like Cochrane Library, PubMed, Scopus etc. and several published articles, and government reports/policy documents with reference to SCD.

Results

A total of 15 original research articles and 2 policy/program documents are included in this review. The review suggests a prevalence of 0.4%–39% studies conducted among medical students, tribal schoolchildren, and tribal adolescents, with diverse screening methodologies.

Conclusion

Diversified prevalence is observed in this review. Several screening methods including dithionite turbidity

test/hemoglobin/high-performance liquid chromatography methods were used to estimate the prevalence, stating the requirement of standardization. This research identified that not only tribal population, but also non-tribal population have the risk of getting affected by SCD. This alarming situation needs comprehensive investigation.

Keywords: Maharashtra, sickle cell, sickle cell diseases**Introduction**

Sickle cell diseases (SCDs) is an evolving public health confrontation, not only in India but also in developed countries of the World [1]. According to the estimation of several studies, between 2010 and 2050, around 14.2 million babies will be born with sickle cell anemia (SCA) [2]. More than 75% of the total global SCD cases are prevalent in Africa continent, while in few parts, the prevalence of sickle cell trait (SCT) (heterozygous) is around 30% [3]. In the year 2006, the World Health Organization recognized SCD as a global public health problem [4]. The reports of published literature warn multiple effects of epidemiological and demographic transitions in low-to-middle-income countries and their consequences for SCD burden.

In India, SCD has been geographically distributed within the central and western regions [5, 6]. The prevalence of sickle cell gene is around 5%–34% in various scheduled

tribes (STs), who are socioeconomically disadvantaged and frequently warrant medical intervention [7]. As per the present prevalence of SCD, there would probably 20 million SCT and 1.9 million SCD patients among the tribal population [8]. In India, it is challenging to determine the burden of SCD, without a gold standard-based screening programs, nationwide reporting system, and/or registries [9]. Maharashtra with 96.87 lakh tribal populations is expected to have at least 11,00,000 SCT and 90,000 SCD patients [10]. To prevent SCD, Maharashtra Arogya Mandal was established in the year 1960. The objective of this study was to determine the prevalence of SCD in several communities and types of screening methods employed in Indian studies.

Material and Methods

An in-depth literature review was performed with the Search Engines like PubMed, ScopeMed, Google Scholar, and Cochrane Library. Several published articles, important government reports, and some policy documents with specific reference to SCD were the literature sources for the current review. We both the authors independently searched for all the potential articles in the above-mentioned search engines, and summarized and cross-checked for duplication and finalized the articles as per the inclusion criteria.

The following keywords were examined in the title or abstract of the research manuscripts: “sickle cell(s),” “sickle cell disease(s),” “sickle cell anemia,” “sickle cell crisis,” “sickle cell disorder(s),” “sickle cell condition(s),” “sickle cell trait(s),” “sickle cell hemoglobin(s),” “prevalence,” “India,” “Maharashtra,” “screening,” “Ante natal screening,” “Programs,” “Policies,” “Control,” and “IEC.” The following filtering criteria were adopted in the electronic search: (i) only studies published between December 1997 and November 2017 were considered, to focus on a description of the current use/issues; (c) and

only original papers and systematic reviews were included in the current review.

Results

Depending on the inclusion criteria set in our review, 15 original articles and 2 policy documents from Maharashtra until November 2017 are considered in this review. In this review, most of the published literature are cross-sectional studies performed on an adolescent population focusing on screening. The methods employed for screening and the documentation of the prevalence was different in different studies. These methodologies ranged from dithionite turbidity test (DTT) followed by electrophoretic test to sophisticated high-performance liquid chromatography (HPLC). Pagrut et al. used DTT as an electrophoretic test and concludes that the prevalence of sickle cell disease among backward classes in Yavatmal district is more especially in schedule caste (SC) in comparison to general or open category with higher prevalence among female subjects [11]. Another study utilized Solubility Test (ST) as screening method followed by cellulose acetate membrane electrophoresis [12].

In a voluntary community screening program, using HPLC methodology, Colah report it may be important to screen for G6PD deficiency in newborn screening programmes for sickle cell disorders [13]. A study by Warghade et al. documented the prevalence of 1.59% for SCD using CE-HPLC [14].

In a study based on camp approach for screening of SCD using DTT among universities, it documented Bhils, Madias, Pawaras, Pardhans and Otkars as the most commonly affected castes [15, 16]. Other prevalence studies among tribal populations of Pombhurna, Chandrapur suggest the prevalence of SCD to be around 5.54% [17]. In Maharashtra, the Korku, Bhills, Gaoli, Gowari and Nihal tribes of Amravati have a high documented prevalence of sickle hemoglobin (HbS) [18].

Another larger community-based study screened 1078 samples of tribal population in Yawatmal, which found that the overall prevalence of SCD in South Maharashtra was 4.14% [19]. Long term evaluation of such efforts and its integration with the existing health system is the need of the hour. Newborn screening program in Maharashtra screened 3448 newborns using HPLC. The SCD babies were followed up clinically and hematologically regularly for 1.5–5 years to describe the course of the disease [20].

Discussion

Most of the published articles aim on screening the knowledge, attitudes and practices or hematological profile of SCD patients. The prevalence of SCD ranged from 0.4% to 39%. Nevertheless, this cannot be generalized since studies have employed diverse methodologies with different approaches and different classification of castes and target population. Moreover, there is no uniformity in the screening test used and its validity. Published studies have employed DTT/HB/HPLC methods to evaluate the prevalence of SCD. Available published findings suggest that both tribal and nontribal populations are predisposed to SCD, however there are no systematic scientific studies to investigate the risk of acquiring SCD among the non-tribal population.

Conclusion

There is a crucial requirement to establish standardization for screening of SCD. A gold standard methodology for screening of SCD needs to be established.

Acknowledgment

The authors are grateful to the Library assistance in literature searching during the initial phase of manuscript.

Financial support and sponsorship

Nil.

References

[1]. Ansong D., Akoto AO., Ocloo D., Ohene-Frempong K. Sick cell disease: management options and challenges

in developing countries. *Mediterr J Hematol Infect Dis*. 2013; 5(1), e2013062. doi: 10.4084/MJHID.2013.062.

[2]. Piel FB., Hay SI., Gupta S., Weatherall DJ et al. Global burden of sickle cell anaemia in children under five, 2010-2050: modelling based on demographics, excess mortality, and interventions. *PLoS Med*. 2013;10(7):e1001484. doi: 10.1371/journal.pmed.1001484.

[3]. Grosse S., Odame I., Atrash H., Amendah D et al. Sick cell Disease in Africa. A Neglected Cause of Early Childhood Mortality *Am J Prev Med*. 2011; 41(6): S398–S405. doi: 10.1016/j.amepre.2011.09.013

[4]. United Nations General Assembly, 2009, Recognition of sickle-cell anaemia as a public health problem.

[5]. Tewari S., Rees D. Morbidity pattern of sickle cell disease in India: A single centre perspective. *Indian J Med Res*. 2013; 138(3): 288–290.

[6]. Saraf S., Molokie R., Nouriaie M., Sable C et al. Differences in the clinical and genotypic presentation of sickle cell disease around the world. *Paediatr Respir Rev*. 2014; 15(1): 4–12.

[7]. Shrikhande AV., Arjunan A., Agarwal A., Dani A et al. Prevalence of the $\beta(S)$ gene among scheduled castes, scheduled tribes and other backward class groups in Central India. *Hemoglobin*. 2014; 38(4):230-5. doi: 10.3109/03630269.2014.931287.

[8]. Balgir S. The spectrum of haemoglobin variants in two scheduled tribes of Sundargarh district in north-western Orissa, India. *Annals of Human Biology*. 2005; 32(5): 560-573

[9]. Balgir S. The Challenge of Haemoglobinopathies in India. *The National Medical Journal of India*. 1999; 12(5): 1-10.

[10]. Sahas B., Goyal R., Yogesh R. Sick cell anemia and morbidity in tribal population of Pombhurna, district Chandrapur, Maharashtra, India. *Innovative Journal of Medical and Health Science*. 2014; 4(6): 169 – 171

- [11] Keshav Pagrut K., Chide P. Screening for the sickle cell gene in Yavatmal District, Maharashtra, India: An approach to a major public health problem. *International Journal of Biomedical and Advance Research*. 2017; 8(02): 50-53.
- [12] Dhumne U., Jawade A. Sickle Cell Anemia and Morbidity in Rural Population of Chandrapur District, Maharashtra, India. *The Anthropologist*. 2011; 13(1): 61-63.
- [13] Colah R., Mukherjee M., Martin S., Ghosh K. Sickle cell disease in tribal populations in India. *Indian J Med Res*. 2015; 141(5): 509–515.
- [14] Warghade S., Britto J., Haryan R., Dalvi T. Prevalence of hemoglobin variants and hemoglobinopathies using cation-exchange high-performance liquid chromatography in central reference laboratory of India: A report of 65779 cases. *Journal of Laboratory Physicians*. 2018; 10(1): 73-79.
- [15] Kate S., Lingojar D. Epidemiology of Sickle Cell Disorder in the State of Maharashtra. *Int J Hum Genet*. 2002; 2(3): 161-167.
- [16] Singh KS. Calcutta, India: Anthropological Survey of India. 1992.
- [17] Lehman H., Catbush M. Sickle cell trait in Southern India. *BMJ*, 1952; 404.
- [18] Zade V., Chede S, Thakre V, Warghat N. The prevalence of sickle cell disease phenotypes and sickle cell gene frequency in some tribals of Melghat forest region of Amravati, Maharashtra (India). *Biosci. Biotech. Res. Comm*. 2011; 4(1): 70-73.
- [19] Patki V. The Prevalence of Sickle cell Disease Phenotypes and Sickle Cell Gene Frequency in Some Tribals of Ghatanji and Kelapur Taluka, Distinct Yavatmal, Maharashtra (India). *International Journal of Scientific and Research Publications*. 2013; 3(5): 1-3.
- [20] Kapoor S., Gupta N. National Newborn Screening Program – Still a Hype or a Hope Now? *Indian Pediatr*. 2013;50(7): 639-43.