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Awareness about Sickle Cell Disorders amongst the Population in Central India – A Cross Sectional Study

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A B S T R A C T

Introduction:Sickle Cell Disorder (SCD), marked by abnormal haemoglobin Hb S, causes rigid, sickle-shaped erythrocytes, blocking circulation and leading to multi-organ damage. Globally, over 3,00,000 infants are born with SCD every year, with India ranking second in prevalence, affecting 20 million people. While advanced nations benefit from neonatal screenings and immunization, low-income regions, including rural India, face high childhood mortality rates (50-80%). Lack of awareness, delayed diagnosis, and inadequate management contribute to elevated mortality, particularly in less-educated rural and tribal areas. This study aims to evaluate public awareness of SCD to address the pressing issue in India.

***Material and Methods*:** This was a cross-sectional study of both known cases of Sickle Cell Disorder (SCD) and non-SCD individuals. A total of 169 consenting individuals were interviewed using a structured validated questionnaire.

***Results*:** We observed lack of awareness regarding Sickle Cell Disorders (SCD) in 40% of the participants, while 60% were found to be aware of its existence.

***Conclusion*:** The study reveals a notable lack of awareness about Sickle Cell Disorder (SCD), including its causes, diagnosis, and treatment. Given the substantial contribution of central India to annual SCD cases, addressing these awareness gaps is imperative. Targeted educational efforts, screenings and awareness campaigns are vital to bridging this knowledge deficit and improving outcomes in the region.

Keywords: Awareness, Sickle Cell Disorders.

INTRODUCTION

Sickle cell disorder (SCD) is a genetic defect of haemoglobin which occurs due to the substitution of valine in place of glutamic acid in the sixth position of the Beta globin chain of haemoglobin due to abnormal haemoglobin Hb S [1, 2]. Sickle cell disorders consist of Sickle Cell Trait, Sickle Cell Anaemia and Sickle Cell diseases (Hb S in combination with normal or a hemoglobinopathy gene). In deoxygenated state, conformational changes caused by Hb S lead to sickling of red blood cells. These sickle-shaped cells are rigid and can block the blood circulation which may lead to progressive multi-organ damage, and thus, increased morbidity and mortality [3, 4].

More than 300,000 infants are born annually worldwide with SCD [5]. Currently, SCD is the commonest single gene defect in the world, most prevalent in sub-Saharan Africa, Mediterranean countries, the western Europe, India and in the Middle East [6]. In India, which has ranked the second most affected country there are more than 20 million SCD patients [7]. In India, the disease is seen mainly in Madhya Pradesh, Orissa, Andhra Pradesh, Vidharbha region of Maharashtra, Kerala (Chetti tribes of Wayanad district) and Tamil Nadu (Nilgiri).

The life expectancy and quality of living of SCD patients can be significantly improved with procedures like neonatal screenings as well as immunization against infections. Prenatal screening is also important. They have found to drastically reduced childhood mortality rate in high-income nations. In such countries, more than 90 per cent of children with SCD survive to adulthood [8]. Such measures are however, not widely implemented, particularly in low income nations, where 50-80 per cent of children with sickle cell disease die even before reaching 5 years of age [9]. The same is true for the less educated, rural and tribal population of India, which take up to around 25% of the country. According to an analysis done in Gujarat, around 20 per cent of infants with sickle cell disease survived only up to two years whereas, 30 per cent children from tribal areas died before they reached adulthood [10].

Our state has launched a screening program for the rural population called "BLOSSOM" (acronym for breast cancer, liver and lifestyle diseases, oral cancer, sickle cell disease, sexually transmitted infections, osteoporosis and malnutrition). Hence, this also emphasizes the importance of awareness about sickle cell disease [11].

It is evident that SCD is a major problem in India. Although various measures to prevent and reduce the occurrence of SCD throughout the world are being implemented, due to lack of awareness in the majority of our population, late diagnosis and improper management systems, the mortality rate of individuals with sickle cell disease in India is high, especially in the rural and tribal areas [12]. Hence, the present study is designed to assess the existing knowledge of the general public regarding SCD.

Objective: To find out the awareness about sickle cell disorders among people residing in central India.

METHODOLOGY

This cross-sectional study involved 169 consenting individuals. The research utilized a structured, validated questionnaire to interview both known cases of Sickle Cell Disorder (SCD) and non-SCD individuals.

Sampling Method: Random sampling method

Statistical Analysis: Results were analysed using STATA version 10.1 (2011).

OBSERVATIONS AND RESULTS

In the study involving 169 participants, 40% demonstrated no awareness of Sickle Cell Disorder (SCD), while 60% were found to be aware of its existence.

Table 1:Age group & sex of respondents

Age in Years	Male	Female	TOTAL
0-10	3	0	**3**
11-20	15	36	**51**
21-30	17	26	**43**
31-40	19	17	**36**
41-50	12	11	**23**
51-60	5	2	**7**
61 & above	4	2	**6**
Total	**75 (44.4%)**	**94 (55.6%)**	**169 (100%)**

Table 2:Source of information about SCD

SOURCE	NUMBER
School/ College	80
Relative/ Friend	25
News paper	5
Social Media	16

Table 3: Respondents replies to what they think causes sickle cell disorder

RESPONDENT'S VIEW	NUMBER
Hereditary	**94**
Don't know	**67**
Others (Infection/ IDA/ High WBC count)	**58**

Table 4: Responses of the participants to questions asked regarding awareness of sickle cell disorder

QUESTION	YES	NO
Awareness about sickle cell disorders	**102**	**67**
Are family members affected by SCD	**11**	**158**
Awareness about blood tests/ screenings to diagnose SCD	**57**	**114**
Awareness about communities prone to SCD	69	100

DISCUSSION

In our study we had a total of 169 participants. The male and female participants were represented by 44.4% and 55.6% respectively (Table 1). 31% of our responders were patients with Sickle Cell Disease (SCD) (including both AS and SS patterns) and 6% individuals had a family member who suffered from SCD.Maximum number (30%) of responders were within the age group of 11 to 20 years. In a similar study in Nigeria, response rate was 83.71%, with male participants in the majority as indicated by 52% of the sample. Their dominant age group was 21 to 30 years (47.90%) [13].

Out of the total participants, 40% were not aware about SCD at all and 60% of the participants were found to be aware of the existence of SCD. In a survey done among antenatal women in Ghana, 10.2% of the respondents answered that they had adequate knowledge about SCD, 48.5% answered they had some knowledge and 41.3% said they knew nothing at all [14].

In our study, when the individuals were asked about their sources of information regarding sickle cell disease (SCD), they provided multiple responses (Table 2). Among the responders, 60% indicated that they first learned about SCD in their School/College environment. Additionally, 20% reported acquiring information about SCD from their relatives or friends. The remaining 20% mentioned obtaining information about SCD from sources such as social media or newspapers.

Level of education was one of the factors influencing the participants' perception towards the disease, this has been demonstrated in other studies as well [15, 16]. Undergraduates 35%, postgraduates 22% and most of the 12th pass individuals had the right information about the cause for SCD. While 54% of the total responders were correctly aware about the hereditary nature of SCD (Table 3), 6% of the participants were mistaken about their knowledge. The misconceptions about the cause of SCD, included factors like infections, iron deficiency anaemia, or high white blood cell count. Similar results were seen in a study undertaken in Benin, West Africa where respondents were not sufficiently aware of the hereditary nature of SCD [17]. Some participants in a Nigerian study wrongly believed that bacterial or viral infections are among the risk factors of sickle cell disease [13]. This is possibly because SCD patients are more prone to bacterial infections [18]. In another study by Agbanusi*et al*., it was seen that even people who were aware of SCD, were not aware of its actual genetic implications since only one out of three obligate carriers (people who have either a parent or a child with SCD) among their participants were aware of their exact trait condition [19]. When asked about the symptoms of SCD, multiple participants correctly listed fatigue, anaemia, multiple frequent blood transfusions, repeated episodes of jaundice, joint pain and swelling [20].

Only 20% of our respondents were aware about the importance of pre-marital genetic screenings. This is comparable to a study done in 2020 in Odisha where 93% of the participants had heard about SCD but about 70% of them had no idea about premarital screening [21]. In a study held amongst Nigerian unmarried adults, most of the study participants (67.1%) suggested that they would prioritize sickle cell screening in their relationships [13].

Among our participants, 33% were aware about the blood tests and screenings done for the diagnosis of SCD and its importance in pregnancy (Table 4). Pregnancy with SCD requires more frequent ANC visits, foetal surveillance, proper nutritional, haematological and emotional support [22, 23, 24, 25]. In a study performed in Gujarat, it was found that increased awareness has caused an increased need for antenatal diagnosis [26]. Other African studies indicate that the expense of attending antenatal clinics in developing countries may be an obstacle, especially in rural areas [27].

Apart from pre-marital and antenatal screenings for SCD, new-born screenings also play an important role in early diagnosis and reduced morbidity of SCD. According to a study performed in 2021, the affected children are generally identified only after they start presenting with symptoms [28]. Some regional neonatal screening programs have been initiated in the last decade [29, 30, 31, 32, 33]. There is need for a national new-born screening program for SCD in India [34].

When asked about the different communities more prone to SCD, 40% of the participants responded with answers like- African/ Scheduled Tribes/ Sindhi community/ Vidarbha population (Table 4). In India, Sickle cell disease (SCD) is indeed more prevalent among certain communities in India, particularly among Scheduled Tribes (ST) (0-35%) and Scheduled Castes (SC) [1]. The tribal groups with a high prevalence of SCD include the Bhils, Madias, Pawaras, Pardhans and Otkars[34]. But contrary to the common earlier belief, it is also seen in non tribal population in central India [35]. It is also common in Vidarbha, Satpura ranges, some parts of Marathawada, Madhya Pradesh, Chhatisgarh, Orissa, Gujarat, Andhra Pradesh and Nilgiri Hills of Tamil Nadu in India [36]. Particularly in Maharashtra, districts like Gadchiroli, Nagpur, Bhandara, Chandrapur, Yeotmal and Nandurbar have more than 5000 cases of SCD [37].

Possible reasons for the current situation include: A substantial lack of knowledge of sickle cell disease (SCD) amongst the participants, indicate a significant gap in education and information dissemination. The study suggests a need for more diverse and accessible channels for disseminating information about SCD since most of the participants were just aware of SCD because of their academic knowledge. This survey also indicates a lack of accurate information and awareness campaigns. The lack of awareness about diagnostic procedures and their importance in managing the disease is concerning.

Potential Solutions for the above cited lack of awareness includes the following:

Comprehensive Education Campaigns: Implement targeted and comprehensive educational programs in schools, colleges, and communities to increase awareness about SCD. Include accurate information about the causes, symptoms, diagnosis, and treatment options of SCD.

Diverse Information Channels: Utilize various information sources such as social media, community events, healthcare centres, and mass media to disseminate information about SCD. Engage influencers, healthcare professionals, and community leaders to spread awareness. Conduct targeted campaigns to debunk myths and misconceptions about SCD causes and transmission.

Tailored Educational Materials: Develop educational materials like pictorial pamphlets, videos in multiple languages and formats accessible to different age groups and educational backgrounds.

Enhanced Healthcare Access: Ensure access to healthcare facilities providing SCD screening and diagnosis, especially in regions with a higher prevalence. This includes educating healthcare professionals to improve early diagnosis and management.

Genetic Counselling and Family Planning: Offer genetic counselling services to individuals and families at risk of carrying the sickle cell gene, emphasizing pre-marital screening for SCD and the importance of knowing one's genotype.

By implementing these measures, there can be a significant improvement in raising awareness, correcting misconceptions, and providing accurate information about sickle cell disease, thereby positively impacting the affected individuals and communities.

CONCLUSION & SUMMARY

Based on the above study, we can conclude that there is a significant gap in education & awareness regarding SCD, its cause, diagnosis and treatment options. Since India, especially central India contributes to a large proportion of SCD cases every year, it is important to overcome these shortcomings in awareness of SCD by conducting SCD targeted programs like:

1) Pre-marital screening
2) National neonatal screening
3) Genetic counselling
4) Awareness programs for public using diverse sources like social media, healthcare professionals and community leaders.

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