RESEARCH ARTICLE

Sickle Cell Anemia – A Societal and Clinical Challenge in a Wardha City

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ABSTRACT

Sickle cell anemia is a hereditary blood disorder that poses significant clinical and societal challenges, predominantly affecting individuals globally. Clinically, it is marked by severe pain episodes, organ damage, heightened infection risk, and limited treatment options. These complications necessitate comprehensive, ongoing medical care. Societal challenges include healthcare disparities, stigma, psychosocial impacts, and barriers to education and employment. Addressing these issues in Wardha city requires improving access to specialized care, enhancing public awareness, investing in research for new treatments, and providing robust support systems for patients and caregivers. A multifaceted approach involving healthcare providers, researchers, policymakers, and communities is essential to effectively manage and mitigate the impacts of this debilitating disease.

Keywords: Heredity, Sickle cell anemia, Treatment.

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INTRODUCTION

Sickle cell anemia is an autosomal recessive syndrome that results from a single amino acid substitution in the beta-hemoglobin molecule. This results in a distortion from the normal biconcave disk shape of the RBC to a crescent or "sickle" shape. Such abnormally shaped cells are less able to deform when passing through the microvasculature and are more prone to hemolysis. Hemolysis of the sickle cells and the resultant inability of the damaged endothelium to release enough nitric oxide to keep the blood vessels dilated predispose to vaso-occlusive crises, which are the clinical hallmarks of sickle cell disease. This will be covered in more detail later in this study and is the root cause of many of the serious consequences associated with sickle cell anemia. At the sixth position in the beta-hemoglobin chain, valine replaces glutamic acid with a single amino acid.[1] The gene will result in sickle cell trait (AS) rather than the disease when inherited from a single parent. Sickle cell trait carriers are often in good health. They are carriers of the defective gene and can pass it on to their children. With one abnormal hemoglobin gene and one normal gene, people with sickle cell trait have mostly normal hemoglobin, in contrast to sickle cell disease. A high level of sickling under certain conditions will occur in people with sickle cell trait or an attack of sickle cell disease. Hemoglobin S turns normal, round RBCs into abnormal, sickle shapes. The cells are fragile and prone to breaking apart.[2] The sickle-shaped cells frequently die after only about 10 to 20 days. The body is unable to swap the lost cells at that rate, leading to a chronic shortage of RBCs.[3,4] This disorder is called anemia, and it can make a person tired or weak. Because sickle cell disease patients have a lower number of RBCs, which are normally rich in iron, it can lead to the development of iron overload.

Signs and Symptoms

Symptoms of sickle cell anemia usually appear at six months of age. They are flexible and vary from person to person. The following are some of the symptoms:

Anaemia

Sickle cells are fragile and finally die. Red blood cells need to be replaced after 120 days on average. Sickle cells, however, frequently run out in 10 to 20 days, leaving a shortage of red blood cells. This is known as anemia. If the red blood cell count is low, the body cannot get adequate oxygen.[4]

Painful episodes

One of the primary indicators of sickle cell anemia is agony crises, which are recurrent bouts of intolerable anguish. Pain rises when sickle-shaped red blood cells block blood flow to the chest, abdomen, and joints. Depending on how bad it is, the pain could last a few hours or several days. Some have scarce or no pain crises in an assumed year. A year, some people have twelve or more. A significant pain crisis requires hospitalization.[5]

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Treatment

- Transplanting stem cells or bone marrow can cure sickle cell disease (SCD). Since these transplants carry a significant risk and can have disastrous adverse effects, they are often only performed on children with severe SCD. For the transplant to be successful, the bone marrow must match precisely. Usually, a sibling or sister would make the perfect donor.[6] Gene therapy is utilized to treat SCD in patients who have had several sickle cell crises and are 12 years of age or older. These cutting-edge treatments employ some of your blood stem cells, and they either have their DNA changed or replaced.
- You will then receive these cells back, and they will be able to produce a healthy kind of hemoglobin.[7] This may lessen SCD crises and other associated consequences.

Therapies that can reduce problems and alleviate symptoms include: Antibiotics to try to keep younger kids from getting sickle cell anemia.

MATERIALS AND METHODS

Study Design

This cross-sectional survey was conducted with the localities of Wardha city. This study was done on people diagnosed with sickle cell anemia to create awareness amongst people regarding sickle cell anemia.[8]

Study Setting

This study was conducted in different areas of Wardha city.

Sample Size

The criteria for the selection of sample size for this survey was in the range of 50 to 60; among these, 54 responses were obtained.

Inclusion Criteria

The people who are aware and have sickle cell anemia were included living in Wardha city, Maharashtra.

Exclusion Criteria

The people who were not aware of sickle cell anemia were excluded.

Questionnaire Development

The questionnaire developed for this survey was made through a literature search in Google Form mode (https://forms.gle/ZjdUbt7cFsAXhxCW6). Two pharmacy professors did the validity testing of the questionnaire, and the required changes were made as per their suggestions. This questionnaire was composed of 19 questions.

Ethical Considerations

The study adhered to ethical guidelines approved by the Ethics Committee of DMIHER, Wardha, Maharashtra, ensuring that it was related to the pharmacy profession.

RESULTS AND DISCUSSION

Table 1 represents the age group in the survey is 15 to 25 years, accounting for 70.4% of the responses. This indicates that the majority of respondents are young adults. The second largest group is the 26 to 40 years age range, with 22.2% of the responses. The 6 to 14 years age group comprises 5.6% of the respondents. The above 40 years age group is minimally represented, with only 1.9%. There are no responses from the 0 to 5 years age group. Males constitute the majority of respondents, with 57.4% of the total responses. Females make up the remaining 42.6% of the responses. This distribution indicates that the survey has a higher number of male participants compared to female participants, with males outnumbering females by a margin of 14.8 percentage points.[9]

Table 1 a: Demographic responses from people (n = 54)

Age (years)	*No. of responses (%)*
0–5	0
6–14	3 (5.6%)
15–25	38 (70.4%)
26–40	12 (22.2%)
Above 40	1 (1.9%)

Table 1 b: Gender-wise responses

Gender	*No. of responses (%)*
Male	31 (57.4%)
Female	23 (42.6%)

Table 2: At the age of diagnosis, sickle cell anemia

Age at diagnosis (Years)	*No. of responses (%)*
Under 1 year	6 (11.1%)
1–5	2 (3.7%)
6–10	8 (14.8%)
11–18	14 (25.9%)
Over 18 years	24 (44.4%)
Rate of severity of symptoms	*No. of responses (%)*
Mild	20 (37%)
Moderate	27 (50%)
Severe	7 (13%)

Table 3: How often do people experience pain crises?

Often experience pain crises	*No. of responses (%)*
Daily	2 (3.7%)
Weekly	4 (7.4%)
Monthly	13 (24.1%)
Yearly	11 (20.4%)
Rarely	24 (44.4%)

Table 2 represents the most common age at diagnosis is over 18 years, with 44.4% of the respondents being diagnosed in this age group. The second most common age group for diagnosis is 11 to 18 years, accounting for 25.9% of the responses. About 6 to 10 years age group comprises 14.8% of the respondents. Under one year age group makes up 11.1% of the responses.[10] The least common age group at diagnosis is 1 to 5 years, with 3.7% of the respondents. This distribution indicates that the majority of diagnoses occur in adulthood (over 18 years),

Table 4: Knowledge and understanding about sickle cell anemia

Questions	*Yes (%)*	*No (%)*	*Unsure (%)*
Family history of sickle cell anemia	6 (11.1)	40 (74.1)	8 (14.8)
Have you encountered challenges in accessing appropriate medical care for sickle cell disease?	23 (42.6)	31 (57.4)	-
Do you feel adequately informed about the available treatment options for sickle cell disease?	26 (48.1)	28 (51.9)	-
Have you experienced discrimination or stigma related to your condition?	21 (40.4)	31 (59.6)	-
Do you participate in any support groups or communities for individuals with sickle cell disease?	21 (38.9)	33 (61.1)	-
Have you ever considered participating in clinical trials or experimental treatments for sickle cell disease?	26 (48.1)	28 (51.9)	-
Have you had genetic counseling or testing related to sickle cell disease?	25 (46.3)	29 (53.7)	-
Do you face any specific challenges in managing sickle cell disease during pregnancy or planning for a family?	18 (33.3)	36 (66.7)	-

followed by adolescence (11–18 years). The early childhood (1–5 years) group has the fewest diagnoses.[11]

Table 2 also represents the severity rate of symptoms of Sickle cell anemia. The majority of responses fall under the "Moderate" category, with 50%. "Mild" symptoms represent 37% of responses, and "Severe" symptoms represent 13%. Table 3 represents the data based on the responses; it appears that the majority of individuals (44.4%) rarely experience pain crises. Following that, 24.1% experience them monthly, 20.4% yearly, 7.4% weekly and only 3.7% daily.[12]

Overall, the data reflects a range of satisfaction levels, suggesting that while some individuals feel well-supported and cared for, others may perceive gaps or shortcomings in the assistance they receive.

Family History

- 11.1% of respondents reported having a family history of sickle cell disease.
- 74.1% indicated no family history.
- 14.8% were unsure about their family history (Table 4).

Access to Care

- 42.6% of respondents said they encountered challenges in accessing appropriate medical care for sickle cell disease.
- 57.4% reported no problems accessing care.[13]

Treatment Awareness

- Nearly half (48.1%) of respondents felt inadequately informed about available treatment options.
- Slightly over half (51.9%) felt they had sufficient information.

Discrimination

- A significant portion (59.6%) of respondents reported experiencing discrimination or stigma related to their condition.
- 40.4% said they hadn't experienced discrimination.[14]

Support Groups

- 61.1% of respondents participated in support groups or communities for individuals with sickle cell disease.
- 38.9% did not participate in such groups.[15]

Clinical Trials

- The interest in clinical trials was almost evenly split, with 48.1% unsure or not interested and 51.9% having considered participation.

Genetic Counseling/Testing

- Over half (53.7%) of respondents had undergone genetic counseling or testing related to sickle cell disease.
- 46.3% had not received genetic counseling or testing.

Pregnancy and Family Planning

- While two-thirds (66.7%) reported no specific challenges managing sickle cell disease during pregnancy or planning a family, a significant minority (33.3%) did face challenges.[16]

CONCLUSION

The study entitled, "Sickle Cell Anemia – A Societal and Clinical Challenge in Wardha city," helps in understanding the societal and clinical realization of this hereditary disorder on-populace. Sickle cell anemia patients experience difficulties and stress in their everyday lives, which makes promoting enhanced healthcare policies and public awareness of this disease very important. The study further showed that different demographical sectors within Wardha city were impacted by the changes in a big way with different levels of health and awareness. The study explored the disparities of patients within the clinical setting in terms of the care they received, as well as the positive and negative imperatives of cure strategies that determine patient results. Add to that, it has a social impact that goes as far as patients' families and even communities at large. The affected persons can experience stigma, costs, and interruption of schooling. Exploratory II lack of knowledge about sickle cell anemia among the general population and lack of education on how to prevent the disease and early symptoms of the disease that can be treated effectively makes it important for educational campaigns to be launched. The authors indicate that early interventions, strengthening primary healthcare, and creating community support programs are the

key recommendations. It is believed that since these problems can be effectively dealt with, the standard of living of human beings with SCA in Wardha city will receive a major boost. Hence, the researchers strongly advocate for policymakers, healthcare providers and the local communities to embrace a model of care involving a range of stakeholders that address the social and clinical issues of SCA.

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