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A NARRATIVE REVIEW ON SICKLE CELL DISEASE

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ABSTRACT

Sickle Cell Disease (SCD) is a chronic hereditary hemoglobinopathy with significant biomedical, psychosocial, and socioeconomic consequences. Despite being one of the world's most common monogenic disorders, SCD remains underdiagnosed and undertreated, especially among tribal and underserved communities in low- and middle-income countries, including India. Health-Related Quality of Life (HRQoL) in individuals with SCD is profoundly influenced by pain crises, comorbidities, access to health services, socioeconomic status, psychological resilience, and family support. This narrative review synthesizes evidence from studies published between 2000 and 2024 to examine determinants of HRQoL, awareness levels, and care practices associated with SCD. Findings highlight persistent gaps in awareness, delayed diagnosis, inadequate access to therapies such as hydroxyurea, and culturally rooted misconceptions that contribute to high morbidity and mortality. Conversely, nurse-led education, newborn screening programs, community outreach, and structured self-management interventions have shown promising results in improving outcomes. The evidence underscores the urgent need for a holistic, integrated approach that combines biomedical treatment with psychosocial support and community-based interventions. Nursing professionals can play a pivotal role in screening, awareness creation, adherence counselling, and facilitating culturally sensitive care—particularly in tribal and rural settings. Strengthening nursing and public health systems is essential to reduce the burden of SCD and enhance the quality of life for affected individuals and families.

Keywords: Sickle Cell Disease, Health-Related Quality of Life, Hydroxyurea, Pain Management, Tribal Populations, Nursing Interventions, Community-Based Care, Self-Management, Newborn Screening, Integrated Care

INTRODUCTION

Sickle Cell Disease (SCD) is one of the most prevalent hereditary blood disorders globally and is caused by a point mutation in the β-globin gene, resulting in the formation of Hemoglobin S. The abnormal haemoglobin polymerizes under low oxygen tension, leading to sickling of erythrocytes, vaso-occlusion, chronic haemolytic anaemia, and multi-organ damage. SCD affects millions worldwide, with the highest burden recorded in sub-Saharan Africa, the Middle East, and tribal belts of India **[1]**.

Although treatments such as hydroxyurea therapy, blood transfusions, and hematopoietic stem cell transplantation have significantly improved survival in high-income countries, these benefits are not uniformly accessible. In low-resource settings—including rural and tribal regions of India—delayed diagnosis, limited healthcare access, socio-cultural beliefs, and poor health-seeking behaviour exacerbate disease progression **[2]**.

Nursing interventions, including community-based screening, structured health education, psychosocial counselling, and treatment adherence support, are vital in improving outcomes. This narrative review synthesizes recent evidence on awareness, management, HRQoL determinants, and the role of nursing in addressing SCD, especially in vulnerable tribal communities **[3]**.

Awareness and Cultural Beliefs

A qualitative study involving traditional healers in six tribal districts of India found that most healers associated SCD symptoms with anemia or malnutrition. Many relied on herbal remedies and ritualistic practices due to lack of biomedical knowledge. Engaging traditional healers in SCD programs was suggested as a culturally aligned strategy for improving awareness **[4]**. Kumar *et al*. (2020) reported that tribal adults in Maharashtra held several misconceptions about Sickle Cell Disease, with 68% attributing it to curses or "bad blood" and 54% believing it to be contagious. Only 22% correctly understood its hereditary nature. These misconceptions contributed to delayed diagnosis and low screening acceptance, highlighting the need for culturally tailored health education **[5]**. Many tribal communities still trust traditional healers, believing that illnesses like pain crises originate from spiritual imbalances or the "evil eye." Public hospitals are often used only when symptoms become severe, underscoring how deeply cultural beliefs guide healthcare decisions **[6]**. Suryawanshi *et al*. (2024) found that awareness of Sickle Cell Disease among school-going adolescents in a tribal district of Maharashtra was extremely low—only 5% of the 394

students knew about SCD, and none understood how it is transmitted **[7]**. The study used a quasi-experimental design in Chhotaudepur district, Gujarat, administering a pretested quantitative questionnaire via household surveys in both formative (1,646 participants) and evaluation (1,631 participants) phases **[8]**. Genetic counselling in tribal Indian communities is often challenged by low literacy levels and deep-rooted socio-cultural beliefs, making acceptance difficult **[9]**. Sickle Cell Disease (SCD) places a heavy psychosocial burden on both patients and their families, extending well beyond the physical manifestations of the illness. Research consistently shows that individuals living with SCD face high levels of psychological distress, stigma, and social disruption, which together deeply impact their quality of life **[10]**. Stigma in Sickle Cell Disease is deeply multifaceted: a systematic review of 27 studies found that stigma affects social relationships, psychological well-being, patient–provider interactions, and care-seeking behaviors. To better quantify this in India, the ICMR developed the ICMR-SCD Stigma Scale (ISSSI) using exploratory and confirmatory factor analysis; their validation study (N ≈ 171 patients, 170 caregivers) resulted in reliable 16-item (patient) and 17-item (caregiver) scales **[11]**. The study aimed to conduct a situational analysis of sickle cell disease (SCD) services in the tribal communities of Andhra Pradesh and Telangana. It explored the burden of HbS carriers, availability of screening programs, community awareness, stigma, and the extent of follow-up services. The study highlighted gaps in accessibility, cultural barriers, and underutilized health services, emphasizing the need for nurse-led, community-focused interventions to improve screening uptake and continuity of care. **[12]** Kato *et al*. (2018) conducted a global translational review, synthesizing findings from clinical trials, molecular research, drug development studies, and international guidelines on sickle cell disease. The authors reviewed evidence on gene therapy, small-molecule drugs, and disease-modifying treatments, comparing scientific advancements with real-world access barriers. **[13]** Patrick *et al*. (2019) conducted a program evaluation of an Indian state-level Sickle Cell Disease (SCD) registry designed to improve follow-up and adherence among tribal populations. Data were collected from registry records, follow-up logs, and patient monitoring reports **[14]**. The BABY HUG trial was a multicenter, randomized, double-blind, placebo-controlled clinical trial conducted on infants aged 9–18 months with sickle cell disease. Participants were randomly assigned to receive either hydroxyurea or placebo for two years. The study evaluated

safety, hematologic response, organ protection, and reduction in early SCD complications. **[15]** Ezenwa *et al*. (2016) conducted a randomized controlled trial involving adolescents and adults with sickle cell disease in the United States. Participants were assigned either to a nurse-delivered intervention that included pain diaries, individualized home pain-management plans, and self-management education, or to usual care. Pain intensity, frequency, and healthcare utilization were measured over the study period using validated pain scales and clinic visit records. **[16]** Madour *et al*. conducted a cross-sectional survey among adults in Western Libya to assess knowledge and attitudes toward Sickle Cell Anemia. Using a structured questionnaire, the study analyzed data with descriptive statistics and chi-square tests to determine associations ($p < 0.05$ considered significant). The results showed low knowledge regarding the genetic inheritance of SCA and common misconceptions about symptoms and causes. **[17]** Jaffer *et al*. conducted a descriptive cross-sectional study among adult SCD patients to assess their knowledge and attitudes toward preventive measures of sickle cell crisis. Using a structured questionnaire, data were analyzed with descriptive statistics and chi-square tests ($p < 0.05$). The study found that most patients had low to moderate knowledge regarding crisis prevention, and education level showed a significant association with knowledge scores **[18]**. a cross-sectional study in the Eastern Province of Saudi Arabia to assess public knowledge and perceptions of Sickle Cell Disease. Data were collected using a structured questionnaire and analyzed using descriptive statistics and chi-square tests ($p < 0.05$) **[19]**. a cross-sectional survey among youths in metropolitan Kano, Nigeria, to assess awareness of Sickle Cell Disease and attitudes toward genotype testing. Data were collected using a structured questionnaire and analyzed with descriptive statistics and chi-square tests ($p < 0.05$). The study found moderate awareness of SCD but poor understanding of genotype implications, with significant associations between education level and awareness scores **[20]**. Adigwe conducted a cross-sectional study among unmarried adults in Abuja, Nigeria, to assess knowledge and awareness of Sickle Cell Disease. Using a structured questionnaire, data were analyzed with descriptive statistics and chi-square tests to determine associations ($p < 0.05$) **[21]**.

Genetic Variability and Screening

Colah, Martin, and Ghosh (2015) documented that the sickle-cell gene (HbS) is widely prevalent across multiple tribal groups in India, with carrier rates ranging from 1% to 40% **[22]**. The cross-sectional

study screened 382 newborns from tribal regions of Central India using HPLC to detect hemoglobinopathies (SCD and thalassemia) and a colorimetric assay to identify G6PD deficiency **[23]**.

Community-Based Interventions

Community-based interventions for sickle cell disease that involve traditional healers, local customs, and culturally adapted education have significantly improved awareness, engagement, and adherence in tribal populations. Quasi-experimental studies report 50–60% increases in knowledge about causes, symptoms, and treatment, using methods like difference-in-differences analysis to show statistical significance. These strategies integrate community trust and cultural relevance to enhance intervention effectiveness **[24]**. Babu *et al*. (ICMR India): "Strengthening health system and community mobilization for sickle cell disease screening and management among tribal populations in India" — quasi-experimental intervention in six tribal-dominated districts covering community mobilization, IEC, capacity building, and screening **[25]**. A retrospective observational design was used, utilizing hospital data collected between 2011 and 2015. The study analysed 10,519 maternal admissions from a tribal block hospital in Gujarat. Data were extracted from obstetric registers and case sheets to compare outcomes in SCD and non-SCD mothers **[26]**. The study aimed to evaluate the effectiveness of Gujarat's state-level Sickle Cell Disease (SCD) control program implemented across tribal districts. The program followed a comprehensive public health model consisting of population screening, dedicated SCD clinics, electronic registries, distribution of hydroxyurea, and structured health education. The concept was to strengthen early detection, continuity of care, and preventive practices through a scalable, nurse-driven model tailored to tribal populations **[27]**.

CONCLUSION

Sickle Cell Disease remains a major public health challenge, particularly in tribal and underserved regions where delayed diagnosis, cultural beliefs, and limited healthcare access worsen disease outcomes. Evidence consistently shows that early interventions—such as hydroxyurea therapy, newborn screening, structured health education, and psychosocial support—significantly reduce complications and improve quality of life. However, these advances are often underutilized in low-resource settings due to stigma, misconceptions, and gaps in service delivery.

Studies from India and global contexts highlight that culturally sensitive, community-engaged strategies are crucial for improving awareness, screening

acceptance, and treatment adherence. Nurse-led models, including registries, home-based counselling. pain management support, and community mobilization, have proven effective in enhancing continuity of care and patient empowerment. Furthermore, integrating traditional healers and local health workers into awareness programs strengthens cultural alignment and improves community trust.

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