Health-Related Quality of Life (HRQoL) in Sickle Cell Disease Patients of Maharashtra, India: A Cross-Sectional Analysis Using WHO-QOL BREF and SF-36

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BACKGROUND & OBJECTIVE

Sickle cell disease (SCD) is a lifelong, multisystem haemoglobinopathy in which clinical indices alone do not fully capture the patient's lived burden. This study characterized HRQoL across physical, psychological, social and environmental dimensions and identified demographic, socioeconomic and clinical determinants in 240 confirmed SCD patients from Maharashtra.

METHODS

- Design: Cross-sectional observational study over 18 months across government medical institutions, PHCs and NGO-supported specialist sickle cell clinics in Maharashtra.
- Participants: Any-age patients with confirmed SS, SC or S-thalassaemia variants by HPLC or molecular genotyping; acute crisis/illness and inability to self-report were exclusion criteria.
- HRQoL instruments: WHO-QOL BREF (4 domains; 0–100) and SF-36 (8 subscales; 0–100).
- Clinical assessment: transfusion history, crisis frequency, medications and healthcare utilization.
- Statistics: JMP Pro 16.0.0; mean ± SD, median/IQR; Shapiro–Wilk and Anderson–Darling; t-test/ANOVA; Pearson and non-parametric sensitivity analyses; two-tailed $p < 0.05$ significant.

COHORT PROFILE & CLINICAL MANAGEMENT

• Male predominance: 53.1%; median age 23 years (range 6–65).

• 43.6% of families earned < Rs. 5,000/month; 63.1% of parents were functionally illiterate.

• 35.8% were students; 21.8% were skilled agricultural labourers.

• Mean age at diagnosis: 14.4 ± 9.6 years — substantial diagnostic delay.

• Mean PRBC transfusion rate: 0.65 ± 1.37 units/year; median 0.

• Mean crises in preceding year: 1.2 ± 2.45; median 0.5 (range 0–20).

• >94% received pharmacological therapy; folic acid and hydroxyurea were predominant; self-reported compliance >90%.

• 71.5% received primary SCD care through NGO-supported specialist clinics; 20.1% attended private hospitals.

INTERPRETATION

• Social Relationships was the highest WHO-QOL domain (68.17), suggesting community/family resilience despite socioeconomic vulnerability.

• SF-36 Role Limitation scores showed an “all-or-nothing” functional pattern: median 0 and IQR 100 for both physical- and emotional-role domains.

• The combination of high medication compliance (>90%) and zero documented prophylaxis coverage points to a systemic care-delivery gap rather than simple patient non-adherence.

• The relatively high Environment score (62.96) may reflect access to NGO-supported specialist clinics; this remains a hypothesis in a cross-sectional study.

POLICY IMPLICATIONS

1. Universal newborn screening with district-level HPLC infrastructure.
2. Integrate penicillin prophylaxis and pneumococcal vaccination into routine SCD preventive care.
3. Scale up hydroxyurea availability and prescribing capacity across government and NGO-supported services.
4. Provide vocational transition/skills programs for patients in physically demanding occupations.
5. Embed psychosocial support, peer groups and social-worker services within SCD care teams.

KEY HRQOL FINDINGS

WHO-QOL BREF mean domain scores (0–100)

SF-36: selected subscale findings

Physical Functioning	**74.48 ± 18.27**	Highest continuous domain
General Health	**49.53 ± 21.22**	Lowest continuous domain
Role—Physical	**31.46; median 0; IQR 100**	Marked bimodality
Role—Emotional	**43.15; median 0; IQR 100**	Marked bimodality

DETERMINANTS OF HRQOL

Crisis frequency was inversely associated with Social Relationships ($r = -0.151$, $p = 0.048$) and Environment ($r = -0.183$, $p = 0.016$). Negative trends for Physical Health ($r = -0.133$, $p = 0.081$) and Psychological Health ($r = -0.132$, $p = 0.084$) did not reach significance.

Occupation showed a clinically meaningful gradient in Physical Health: unskilled manual labourers 45.6 vs students 57.0 and semi-skilled agricultural workers 63.0 (ANOVA $p = 0.111$).

Gender was not associated with Physical Health (males 57.1 vs females 57.7; $p = 0.815$).

Parental literacy was significantly associated with Physical Health ($p = 0.037$).

THE PREVENTION GAP

100% of patients with documented preventive-care data (179/179) reported no prophylactic antibiotics, no pneumococcal vaccination and no other SCD-specific preventive care.

LIMITATIONS & CONCLUSION

Limitations: Cross-sectional design limits causal inference; self-reported crisis counts may introduce recall bias; NGO-clinic over-representation may limit generalizability; no healthy control group was included.

CONCLUSION: This 240-patient Maharashtra cohort demonstrates moderate overall HRQoL with three defining features—social resilience, marked functional dichotomy during illness episodes, and a complete documented gap in prophylactic preventive care. Improving outcomes requires a shift from treating the haemoglobinopathy alone to integrated prevention, clinical care, vocational support, health literacy and psychosocial care.

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n = 240	24.2 ± 9.4 y	18 months	0 / 179
CONFIRMED SCD PATIENTS	MEAN AGE	STUDY PERIOD	PROPHYLAXIS COVERAGE