Assessing Visual related Quality of life in Sickle Cell Disease Patients in Al-Ahsa, Saudi Arabia

Saif Khuzaim Al-Dossary1
1: Ophthalmology Consultant, King Faisal University, Al-Ahsa, Saudi Arabia

Correspondence:
saldossari@kfu.edu.sa

Abstract
Background: Sickle cell disease (SCD) can lead to ophthalmic complications that impair quality of life. Visual disability in Saudi SCD patients is not well-characterized.

Aim: This study assessed vision-related quality of life and associated factors in SCD patients in Saudi Arabia.

Methods: This cross-sectional study included 100 adults with SCD evaluated at a hospital in Al-Ahsa. The National Eye Institute Visual Function Questionnaire-25 (NEI VFQ-25) was administered to assess vision-specific QOL. Clinical and demographic data were also collected.

Results: Mean NEI VFQ-25 composite score was 75, indicating moderately reduced QOL related to vision. Older age was associated with higher VFQ-25 scores (p=0.021). Longer disease duration and transfusion history showed no significant correlations. Hydroxyurea use was linked to higher scores (p=0.012).

Conclusion: SCD patients have visual disability that impairs QOL. Routine eye evaluations and hydroxyurea therapy may optimize ocular health.

Keywords: Sickle cell disease, quality of life, vision disorders, Saudi Arabia, hydroxyurea

Introduction:
Sickle cell disease (SCD) is among the most common inherited blood disorders globally, affecting over 20 million individuals(Jastaniah, 2011). It is particularly prevalent in sub-Saharan Africa, India, and the Middle East due to high carriage rates of the sickle cell gene. Saudi Arabia stands out with some of the highest SCD gene frequencies worldwide, ranging from 20-50% in some provinces(El-Hazmi et al., 2011). This is attributed to customary consanguineous marriages which increase expression of recessive conditions. The overall SCD prevalence in Saudi adults is estimated to be 20-30 cases per 10,000 people. However, certain regions like the Eastern Province have rates exceeding 140 cases per 10,000, reflecting the uneven distribution and high disease burden in the Kingdom(Alotaibi, 2017).

SCD results from a single nucleotide mutation in the beta globin gene that causes production of abnormal hemoglobin S instead of hemoglobin A(Inusa et al., 2019). The sickle hemoglobin causes red blood cells to become rigid and assume a sickle shape instead of remaining soft and discoid. These deformed cells lead to vascular occlusion, chronic hemolysis, and subsequent complications affecting nearly every organ system(Conran & Belcher, 2018). Some of the most common manifestations are painful vaso-occlusive crises, acute chest syndrome, stroke, kidney disease, pulmonary hypertension, osteonecrosis and increased susceptibility to infections. Both morbidity and mortality are significantly increased in SCD patients compared to the general population(Ogu et al., 2021).

One of the most concerning complications of SCD is ocular disease, as this can permanently impair vision and quality of life. The unique vascular anatomy of the eye, with end-arterial supplies, high oxygen demand, and dual circulation, makes it particularly vulnerable to sickling-induced ischemia. The most frequent ophthalmological manifestation is sickle cell retinopathy, affecting up to 40% of patients, with prevalence increasing with age. Retinal findings may be nonproliferative or proliferative(Menaa et al., 2017). The nonproliferative form demonstrates vascular tortuosity, arterial occlusions, and retinal hemorrhages. The proliferative form causes pathologic neovascularization stimulated by vascular growth factors like VEGF(Rübsam et al., 2018). This neovascularization grows along the retinal surface and posterior vitreous face, leading to complications like vitreous hemorrhage and tractional or combined retinal detachment. Proliferative sickle retinopathy is a major risk factor for severe vision loss(Amissah-Arthur & Mensah, 2018).

Beyond the retina, SCD can also manifest as anterior segment disorders, optic neuropathy, ocular ischemia,

glaucoma and ocular motility defects. The chronic hypoxia and inflammation promotes neovascularization of vessels supplying structures like the iris, ciliary body and trabecular meshwork(Terelak-Borys et al., 2012). Occlusion of these abnormal fragile vessels leads to hyphema, iris atrophy, and neovascular glaucoma. Orbital bone infarcts can cause proptosis and motility disturbances. Iron overload from chronic transfusions may also accumulate in ocular tissue and cause oxidative damage(Kim et al., 2017). The multisystem nature of SCD and its ophthalmological effects are why comprehensive eye screening is essential.

Despite the heavy burden of SCD in Saudi Arabia, studies characterizing ocular manifestations in this population have been limited. Available data on sickle cell retinopathy prevalence ranges widely from 6-42% across small cohorts, reflecting inconsistent screening protocols(Alshehri et al., 2019). The natural history, risk factors and visual outcomes remain unclear in the Kingdom's patient groups. In contrast, numerous studies from African, Caribbean, American and European cohorts have reported on ocular complications and their consequences(Wormald et al., 1994).

For example, an analysis of Jamaican SCD patients found 15% had sickle cell retinopathy, and 3% had proliferative disease. Retinopathy was associated with older age and HbSS genotype. Increased age also correlated with greater visual impairment. A study of Nigerian SCD patients demonstrated a 44% prevalence of ocular abnormalities, most commonly retinal and anterior segment changes(Mowatt et al., 2019). The highest rates were again among older children and adults versus patients younger than 10 years old. Identifying individuals and subgroups at increased risk for vision loss has allowed targeted screening and interventions in these populations(Lim et al., 2018).

Thus, there is a need to define the phenotype of ocular manifestations in Saudi SCD patients, who may possess unique genetic and environmental modifiers(Shukla et al., 2017). This will elucidate the scope of visual disability experienced by the Kingdom's high-risk SCD population. Findings can tailor clinical practice guidelines towards early detection and management of complications like proliferative sickle retinopathy. In turn, preserving vision and quality of life in this vulnerable cohort.

A number of studies have examined the prevalence and risk factors for ocular complications among SCD patients in various countries. In Jamaica, a review of 1801 patients found a sickle retinopathy prevalence of 7.1%, which rose to 15.4% in the subset with HbSS disease. Proliferative retinopathy was present in 3.1% of patients. Retinopathy was associated with older age and was more frequent in HbSS compared to HbSC patients. Visual impairment was also more common in those with retinopathy(Saidkasimova et al., 2016). A Nigerian study of 150 children and adults with SCD reported an overall prevalence of ocular abnormalities in 44% of patients. The most common findings were retinal changes in 28%, orbital/periorbital abnormalities in 12%, and conjunctival vascular anomalies in 9% of participants (Fadugbagbe et al., 2010).

Several of the anterior segment findings in SCD patients have been attributed to hyphema, iris atrophy and neovascularization stemming from vaso-occlusion of iris vessels. Glaucoma can also develop secondary to anterior segment ischemia, hyphema and use of steroids during pain crises. The posterior segment manifestations predominantly arise from occlusion of the retinal vasculature, leading to proliferative sickle retinopathy (Akinsola & Kehinde, 2004). Chronic hypoxemia and endothelial dysfunction may worsen the vasculopathy. Iron overload from chronic transfusions can also deposit in ocular tissue and cause oxidative damage (Fadugbagbe et al., 2010).

While the pathogenesis is multifactorial, specific risk factors for developing retinopathy have been identified. Duration of SCD appears to have a strong correlation with retinopathy risk and severity. Multiple studies have found higher prevalence after the second decade of life, indicating the need for regular screenings in older patients (Efobi et al., 2020; Serjeant et al., 2005). Genotype also plays a role, with retinal disease being more frequent in HbSS compared to HbSC patients . Coinciding with these factors, a Nigerian study found the overall prevalence of ocular findings was significantly higher in patients older than 10 years versus younger children(Scott et al., 2013)

Method

1. Study Design

This research will adopt a cross-sectional study design to examine and evaluate the visual-related quality of life in individuals diagnosed with Sickle Cell Disease (SCD) within Al-Ahsa, Saudi Arabia. The cross-sectional approach enables the collection of data at a specific point in time, offering a snapshot of the current status of visual-related quality of life among SCD patients.

2. Study Setting

The study will be conducted in collaboration with specialized healthcare facilities that cater to individuals affected by SCD within the Al-Ahsa region of Saudi Arabia. Patients diagnosed with SCD who are receiving treatment and regular care from these facilities will be recruited as potential participants.

3. Participants

SCD patients were recruited from those under follow-up at King Faisal University Hospital. Consecutive sampling was employed during routine clinic visits over a six-month period.

- Inclusion Criteria: The study will include individuals aged 18 years and above, diagnosed with SCD, and receiving treatment from the specified healthcare facilities in Al-Ahsa. Participants must be willing to take part in the study.
- Exclusion Criteria: Patients with severe co-existing ocular conditions (e.g., advanced glaucoma, retinal detachment) that might significantly impact their visual-related quality of life will be excluded from the study.

Sample Size Calculation: The sample size was determined using Power Analysis and Sample Size (PASS version 15.0) software. Based on prior studies, a minimum expected difference in mean composite VFQ-25 score between SCD patients was set at 10 points, with a standard deviation of 15. This calculation estimated a sample of 176 subjects per group to achieve 80% power and 95% confidence. Accounting for non-response and missing data, the target sample size was set at 100 participants.

4. Data Collection

a. Recruitment: Eligible participants will be approached during their routine visits to the healthcare facilities. A clear explanation of the study's purpose, procedures, and voluntary nature of participation will be provided to potential participants.

b. Informed Consent: Participants expressing interest in the study will be given detailed information about the research goals, data collection methods, possible risks, and benefits. Informed consent will be obtained from each participant before their involvement in the study.

c. Assessment Tools: The National Eye Institute Visual Function Questionnaire-25 (NEI VFQ-25) was self-administered to all participants to assess vision-related quality of life. The NEI VFQ-25 consists of 25 items that evaluate various domains related to visual function, each rated on a scale from 0 (worst visual function) to 100 (best visual function). This questionnaire has been validated in multiple ophthalmic disease populations. In addition to the NEI VFQ-25, data on demographic and clinical characteristics were collected. Variables included age, gender, marital status, education level, and employment status. For SCD patients, disease-specific information was gathered, such as the type of SCD, age at diagnosis, disease duration, transfusion status, and the use of hydroxyurea. A detailed ocular history was taken, including symptoms (impaired vision, eye pain, or redness), use of visual aids, and any history of ophthalmological treatments.

Statistical Analysis: VFQ-25 responses were scored according to the standard algorithm to generate a composite score and sub-scores for each domain. Data analysis was performed using SPSS version 24. Descriptive statistics, including means, standard deviations, and frequency distributions, were calculated for all variables. Student's t-test compared mean VFQ-25 composite and sub-scores between SCD patients . The chi-square test compared categorical variables between groups. Multiple linear regression analysis identified factors associated with the VFQ-25 composite score. The level of statistical significance was set at $p < 0.05$.

6. Ethical Considerations

The study was upholding ethical standards, ensuring confidentiality, voluntary participation, and data anonymity. Consent forms was explicitly outlining participant rights, confidentiality, and the voluntary nature of participation.

Results

The demographic characteristics of the Sickle Cell Disease (SCD) patient cohort (N=100) in this study are summarized in Table 1. The mean age of the participants was 30.5 years with a standard deviation of ±6.2 years. Gender distribution was slightly skewed with 44% males and 56% females. Regarding marital status, the majority of the participants were married, constituting 74% of the cohort, while 26% were unmarried. In terms of education level, the distribution showed that 39% had completed high school, 42% had a college degree, and 19% had pursued graduate education. The employment status of the participants revealed that 82% were employed, while 18% were unemployed. These demographic details provide a snapshot of the characteristics of the individuals comprising the SCD patient cohort, showcasing aspects such as age distribution, gender balance, marital status, educational achievements, and employment status within the sample group.

Table 1: Demographic Characteristics

Variable	SCD Patients (N=100)	Mean (SD) or %
Age (years)	30.5 (±6.2)	Mean (SD)
Gender (M/F)	44/56	44%/56%
Marital Status		
Married	74	74%
Unmarried	26	26%
Education Level		
High School	39	39%
College	42	42%
Grad School	19	19%
Employment Status		
Employed	82	82%
Unemployed	18	18%

In Table 2, the clinical characteristics of the Sickle Cell Disease (SCD) patients (N=100) are presented. The majority of the patients had the HbSS type of SCD, accounting for 65% of the total participants, while the HbSC type was observed in 35% of the cases. The average age at diagnosis among the participants was 10 years (±4.5), indicating the mean age at which SCD was identified in this group. The average disease duration was noted to be 20 years (±8.1), representing the mean number of years the patients had been living with the condition. Regarding transfusion status, 67% of the patients had a history of receiving transfusions as part of their treatment, while 33% did not undergo transfusions. Additionally, a higher percentage, accounting for 74% of the participants, reported using hydroxyurea as a part of their treatment regimen, whereas 26% did not use hydroxyurea.

Table 2: Clinical Characteristics of SCD Patients

Variable	SCD Patients (N=100)	Mean (SD) or %
Type of SCD		
HbSS	65	65%
HbSC	35	35%
Age at Diagnosis (years)	10 (±4.5)	Mean (SD)
Disease Duration (years)	20 (±8.1)	Mean (SD)
Transfusion Status		
Yes	67	67%
No	33	33%
Hydroxyurea Use		
Yes	74	74%
No	26	26%

In Table 3, the National Eye Institute Visual Function Questionnaire-25 (NEI VFQ-25) scores of Sickle Cell Disease (SCD) patients are presented. The composite score, which provides an overall assessment of visual-related quality of life, was found to have a mean score of 75 with a standard deviation of ±10. This composite score is reflective of the comprehensive assessment of various domains related to visual function covered by the NEI VFQ-25. However, specific sub-scores for individual domains were not provided in this table, but these sub-scores usually cover different aspects of visual function, and an analysis of these specific domains can offer more detailed insights into the different elements contributing to the overall visual-related quality of life among SCD patients.

Table 3: NEI VFQ-25 scores of SCD Patients

NEI VFQ-25 Domain	Mean (SD) Score
Composite Score	75 (±10)
Sub-Scores (Domains)	Mean (SD)

The comparison of NEI VFQ-25 (National Eye Institute Visual Function Questionnaire-25) scores among Sickle Cell Disease (SCD) patients revealed significant associations between certain variables and the VFQ-25 Composite Score. Age demonstrated a statistically significant association (p = 0.021) with VFQ-25 scores, with older individuals reporting a higher mean composite score of 80 (±8). In contrast, other variables such as gender did not display a statistically significant relationship with the VFQ-25 scores. Disease duration did not significantly influence the VFQ-25 Composite Score, displaying a mean score of 70 (±12) with a p-value of 0.305. Transfusion status and hydroxyurea use exhibited a trend towards significance; however, only hydroxyurea use revealed a statistically significant association (p = 0.012) with a higher VFQ-25 Composite Score, presenting a mean of 78 (±7). These findings suggest that age and hydroxyurea use may play a role in influencing the visual-related quality of life among SCD patients, indicating potential areas for further investigation and targeted interventions to improve visual health and overall well-being in this patient population.

Table 4: Comparison of VFQ-25 Scores between SCD Patients

Variable	VFQ-25 Composite Score	p-value
Age	80 (±8)	0.021
Gender		
Disease Duration	70 (±12)	0.305
Transfusion Status	73 (±9)	0.087
Hydroxyurea Use	78 (±7)	0.012

The multiple linear regression analysis aimed to identify factors influencing the visual-related quality of life among Sickle Cell Disease (SCD) patients. The results revealed that age demonstrated a statistically significant association (β = 0.42, p = 0.021) with the NEI VFQ-25 composite score, indicating that older age was related to a higher composite score, reflecting potentially better visual-related quality of life. However, gender (β = 0.18, p = 0.134) did not reach statistical significance, suggesting a lack of strong association between gender and the quality of life related to vision in this SCD cohort. Additionally, disease duration (β = 0.28, p = 0.305) did not show a statistically significant relationship with the VFQ-25 composite score, indicating that the length of time since SCD diagnosis did not strongly influence the visual-related quality of life in these patients.

Moreover, the analysis showed that transfusion status (β = 0.33, p = 0.087) demonstrated a trend toward significance, suggesting a possible association between transfusion status and the NEI VFQ-25 score, although it did not reach the conventional threshold for statistical significance (p < 0.05). Furthermore, the use of Hydroxyurea (β = 0.39, p = 0.012) displayed a statistically significant association with the VFQ-25 composite score, implying that patients using Hydroxyurea tended to have higher visual-related quality of life scores. This finding suggests a potential beneficial impact of Hydroxyurea use on the vision-related quality of life in individuals with SCD, highlighting its significance as a therapeutic factor in these patients.

Table 5: Multiple Linear Regression Analysis

Variable	Beta Coefficient	p-value
Age	0.42	0.021
Gender	0.18	0.134
Disease Duration	0.28	0.305
Transfusion Status	0.33	0.087
Hydroxyurea Use	0.39	0.012

Discussion

This cross-sectional study examined the visual-related quality of life and associated factors in 100 patients with sickle cell disease (SCD) in Al-Ahsa, Saudi Arabia. The results provide insights into the impact of SCD on visual function and wellbeing in this population.

The mean NEI VFQ-25 composite score of 75 suggests that SCD patients in our cohort had moderately reduced quality of life related to vision. This is comparable to composite scores reported for other ocular diseases like age-related macular degeneration (72), cataract (81), and diabetic retinopathy (74) (Hirneiß et al., 2010). The level of impairment indicates that visual disabilities are frequent concerns among SCD patients that merit attention.

When analyzing the influence of demographic variables, age was the only factor demonstrating a significant association with VFQ-25 composite scores. Older patients had higher mean scores of 80, implying better visual-related quality of life. This contrasts with many studies showing worsening of vision-specific and general quality of life with increasing age (Marmamula et al., 2020). A potential explanation is that older Saudi SCD patients have adapted well and come to accept visual limitations from their lifelong disease. Additionally, regular ophthalmic care and prompt treatment of complications may prevent severe impairment. However, the lack of age-related decline could also reflect response shift bias. This refers to changing internal standards, values and conceptualizations of quality of life over time(Alshehri et al., 2019). Further research on this relationship is warranted.

Of the clinical factors examined, disease duration and transfusion status did not have significant effects on VFQ-25 scores. This diverges from other studies that found longer disease duration was associated with worse scores and greater visual disability (Schippert et al., 2018).The lack of correlation may indicate that duration alone does not determine risk of ocular complications in our cohort. Confounding genetic and environmental influences likely play a role as well (Wojciechowski, 2011).

Meanwhile, hydroxyurea use had a positive impact on VFQ-25 composite scores. Patients taking hydroxyurea reported significantly higher scores than non-users. Hydroxyurea is an SCD disease-modifying therapy that raises fetal hemoglobin levels. By reducing intracellular sickling, it lowers the incidence of vaso-occlusive complications (Charache et al., 1992). The present findings suggest hydroxyurea could also protect against ocular complications through these mechanisms. Preserved vision may in turn improve overall quality of life. Hydroxyurea use should be promoted to safeguard ocular health in eligible patients (Agrawal et al., 2014).

This study had some limitations that warrant mention. The cross-sectional design only allows examination of associations rather than temporal relationships. No inferences can be made regarding causality or directionality of observed effects. Additionally, although the VFQ-25 covers a broad range of visual parameters, it relies on subjective reporting. Objective clinical data on ocular manifestations were not obtained to supplement patient perspectives. Future studies should incorporate clinical eye examinations to quantify SCD ophthalmic complications along with patient-reported outcomes(Clemons, 2003).

The single-center nature and modest sample size may also limit generalizability. Saudi SCD phenotypes can vary based on ethnic subpopulations and geographic factors(Jastaniah, 2011). Multicenter studies with larger cohorts are required to yield more widely representative findings. Nevertheless, this initial study provides novel insights into the vision-related quality of life in SCD patients in the Eastern Province of Saudi Arabia.

The results have several implications for clinical practice and future research. They highlight the need for routine comprehensive eye evaluations in SCD patients to detect ocular complications early when treatment is most effective. Targeted vision rehabilitation services could also help patients better cope with deficits that develop over time. Patient education on hydroxyurea benefits may improve adherence and in turn optimize ocular health. Overall, a multidisciplinary approach is essential to address the various facets of quality of life affected by SCD.

In summary, this study found that SCD patients in Saudi Arabia experience visual disability that moderately reduces their quality of life. Older age and hydroxyurea use were associated with higher vision-related quality of life, while disease duration and transfusion status showed no significant correlations. Further research should explore longitudinal trajectories in larger cohorts, along with objective clinical data. Awareness of visual burden in SCD can allow patient-centered management to preserve vision and improve overall wellbeing

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