

# Vision Quality Assessment in Patients with Sickle Cell Disease Versus Normal Population in Al-Ahsa, Saudi Arabia

Abdullah Fuad Al-Naim<sup>1</sup>, Abdullah Hisham Al-Mulla<sup>1</sup>, Ibrahim Abdulhakim Al-Shikmubarak<sup>1</sup>, Khaled Saleh Al-Mulhim<sup>1</sup>, Saif Khuzaim Al-Dossary<sup>2</sup>

<sup>1</sup>Medical intern, College of Medicine, King Faisal University, Al-Ahsa, Saudi Arabia, <sup>2</sup>Ophthalmology Consultant, King Faisal University, Al-Ahsa, Saudi Arabia

## Abstract

**Introduction:** Sickle cell disease (SCD) is the most common genetic disease worldwide. In Saudi Arabia, the prevalence is estimated to be 4.2% in the adult population. Retinopathy is a well-known complication of SCD. Therefore, it is important to assess the quality of vision in SCD patients and compare them with the normal population.

**Purpose:** The aim of this study is to assess the vision of SCD patients, and compare them with the normal healthy individuals in Al-Ahsa city, Saudi Arabia.

**Materials and Methods:** A questionnaire based study was conducted in Al-Ahsa city from June to August 2017. The sample size is 402. We included SCD patients who don't have any preexisting ocular problems, from the 18 year old and above. The data collection comprised Age, gender, marital status, educational level, time of diagnosis, presence of eye symptoms. The data was analyzed by SPSS version 24 and the p value of <0.05 was considered a statistically significant.

**Results:** A total of 402 individuals in Al-Hasa region were included in our study. 192 (47.8%) of them are known case of sickle cell disease. The minimum age of the participants was 18 and the maximum age was 42. Primary educational level was found in 46 (11.5%), 183 (45.5%) had secondary, and 173 (43%) had the academic educational level. The mean visual questionnaire score was  $6.2 \pm 1.8$ . The data results showed that 150 (37.3%) of the respondents were found to have poor general vision score, 84 (20.9%) of them were known case of SCD. Conclusion: The study concluded that there is no significant relationship between sickle cell disease and vision disabilities. And we recommend other types of studies to be conducted in this important field.

**Key words:** Al-Ahsa, Saudi Arabia, Sickle cell disease, Quality, Vision

## INTRODUCTION

Sickle cell disease (SCD) is a genetic blood disease characterized by presence of abnormal hemoglobin's production and associated with high morbidity and mortality. It is the most common genetic disease worldwide

and in Africans American blacks and middle eastern people in particular<sup>(1)</sup>

It is responsible for approximately annual 113,000 hospitalizations in united states<sup>(2)</sup> and with prevalence estimated to be more than 70,000 in that country<sup>(3)</sup>.

In Saudi Arabia, Information about the prevalence of SCD is patchy and not equal between the regions in the country, but studies have reported that SCD is a relatively common genetic disorder in this part of the world<sup>(4) (5)</sup>.

Previous studies in the country showed that the prevalence of the sickle cell gene in the adult population is 4.2%

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**Corresponding Author:** Abdullah Fuad Alnaim, Huffof, Al-Ahsa, Saudi Arabia. Phone: 00966506039332.  
E-mail: dr.ebda31@hotmail.com

for sickle-cell trait and 0.26% for SCD, with the highest prevalence noted in the Eastern province (approximately 17% for sickle-cell trait and 1.2% for SCD) <sup>(6)</sup>

Sickling of the erythrocyte (RBC) can occlude the blood vessels, decreasing the blood supply to many organs and eventually can damage them <sup>(7)</sup>. One of these organs is the eye leading to many eye complications and blindness in advanced cases <sup>(8)</sup>.

It is well-known that SCD can occlude the blood vessels that supply the retina, causing some sorts of retinopathy <sup>(9)</sup>.

Previous study in our country showed that Peripheral retinal vascular changes were common in sickle cell patients suggesting the strong relationship with the disease <sup>(10)</sup>.

To the best of our knowledge, no study in our region was done before to assess the quality of vision in patients with SCD Making it important to do this kind of study.

### Aim and Objectives of Study

The aim of this study is to assess the quality of vision in people suffering from sickle cell disease and compare them with the normal healthy individuals in Al-Ahsa city of Saudi Arabia

## MATERIALS AND METHODS

A retrospective cross sectional questionnaire based study was conducted from June to August 2017. This research was approved by research ethical committee (REC) in the college of medicine in King Faisal University. The visual function questionnaire-25 (VFQ-25) which was validated by national eye institute was used in this study to assess the quality of vision in two different groups. The first group involves patients suffering from sickle cell disease, and the other group was normal individuals. These groups were chosen randomly. The inclusion criteria involved sickle cell patients who don't have any preexisting ocular diseases and age from 18 years old and above. The researchers have done the questionnaire on 402 individuals. The data collection comprised Age, gender, marital status, educational level, time of diagnosis, and presence of eye symptoms. Data analysis was done by using SPSS program version 24. Descriptive statistics of all variables were obtained to evaluate the data (by chi-square test). The value of  $p < 0.05$  was considered a statistically significant.

## RESULTS

A total of 402 individuals in Al-Ahsa region were included in our study. 192 (47.8%) of them are known case of sickle cell disease (SCD).

The minimum age of the participants was 18 and the maximum age was 62. More than half of them are in the age group (20-40). 199 (49.5%) of them are males.

Primary educational level was found in 46 (11.5%), 183 (45.5%) had secondary, and 173 (43%) had the academic educational level. The demographic characteristics are shown in the Table 1.

Regarding the marital status, 186(46.3%) of them are single, 208(51.7%) are married, and 8(2%) are divorced. The mean visual questionnaire score was  $6.2 \pm 1.8$ . The data results showed that 150 (37.3%) of the respondents were found to have poor general vision score, 84 (20.9%) of them were known case of SCD. On the other hand, 252 (62.7%) were found to have good score, less than half of them [108 (26.9%)] were SCD patients.

The age group  $>40$  had the lowest score in vision assessment (3.2) compared to the age group (20-40) mean score (6.1).

As shown in table 2, around 66% of the respondents of the questionnaire assessed themselves as healthy and have healthy lifestyle. In addition, 324 (81.6%) reported that their eyesight is considered to be healthy. The percentage was slightly much higher in the unaffected individuals (54%) compared the SCD patients (46%). A significant number of the respondents, 295 (73.4%) have worries about their eyesight even though they don't have eye disease at the present time. No significant difference was found between the 2 groups of the study. 26 (13.5%) of the SCD group experienced some eye symptoms like eye pain, burning or itching. the number in the non-SCD group was 32 (15.2%). As shown in Table 3, 45 (23.4%) of the SCD

**Table 1: Sociodemographic characteristics of the respondents**

Demographic characteristics (N=402)	#	%
Age		
<20	116	28.9
20-40	223	55.5
>40	63	15.6
Gender		
Male	199	49.5
Female	203	50.5
Health status		
Sickler	192	47.8
Non-sickler	210	52.2
Marital status		
Single	186	46.3
Married	208	51.7
Divorced	8	2%
Educational level		
Primary	46	11.5
Secondary	183	45.5
Academic	173	43

**Table 2: The responses of different statements related to general health and vision**

Statement	Excellent	Good	Fair	Poor	Very poor
In general, would you say your overall health is	No. 164 (40.7%)	No. 102 (25.3%)	No. 51 (12.9%)	No. 70 (17.4%)	No. 15 (3.7%)
At the present time, would you say your eyesight using both eyes is	Excellent No. 302 (75.2%)	Good No. 22 (5.4%)	Fair No. 5 (1.2%)	Poor No. 43 (10.7%)	Very poor No. 30 (7.5%)
How much of the time do you worry about your eyesight	All the time 118 (29.4)	Most of the time 177 (44)	Sometimes 92 (22.9)	Little 4 (1)	None 11 (2.7)
How much pain or discomfort have you had in and around your eyes (for example, burning, itching, or aching)?	All the time 65 (16.2)	Most of the time 52 (12.9)	Sometimes 139 (34.6)	Little 44 (10.9)	None 102 (25.4)
Would you say it is					

**Table 3: The responses of different statements related to difficulties with activities**

Statement	No difficulty	Little	Moderate	Severe	Extreme
How much difficulty do you have reading ordinary print in newspapers? Would you say you have	300 (74.6%)	61 (15.2%)	12 (3%)	29 (7.3%)	0(0%)
How much difficulty do you have doing work or hobbies that require you to see well up close, such as cooking, sewing, fixing things around the house, or using hand tools? Would you say	No difficulty 282 (70.1%)	Little 97 (24.1%)	Moderate 15 (3.7%)	Severe 7 (1.7%)	Extreme 1 (0.2%)
Because of your eyesight, how much difficulty do you have finding something on a crowded shelf	No difficulty 230 (57.2%)	Little 129 (32.1%)	Moderate 20 (5%)	Severe 18 (4.5%)	Extreme 5 (1.2%)
How much difficulty do you have reading street signs or the names of stores	No difficulty 66 (16.3%)	Little 104 (25.9%)	Moderate 211 (52.4%)	Severe 18 (4.4%)	Extreme 3 (1%)
Because of your eyesight, how much difficulty do you have going down steps, stairs, or curbs in dim light or at night	No difficulty 274 (68.2%)	Little 99 (24.6%)	Moderate 12 (3%)	Severe 9 (2.2%)	Extreme 8 (2%)
Because of your eyesight, how much difficulty do you have picking out and matching your own clothes	No difficulty 332 (82.6%)	Little 20 (5%)	Moderate 36 (9%)	Severe 10 (2.4%)	Extreme 4 (1%)

**Table 4: Impact of sickle cell disease on the vision**

	Presence of SCD		Total
	Affected	Not affected	
Valid			
Poor general vision score			
Count	84	66	150
% of total	20.9	16.4	37.3
Good general vision score			
Count	108	144	252
% of total	26.9	35.8	62.7
Total			
Count	192	210	402
% of total	47.8	52.2	100

group had some difficulties in reading, while the result in the non-SCD group was 28 (13.3%). 43 (10.7%) of the respondents have difficulties in finding something on a crowded shelf, 67.4% of the number mentioned above (29) are SCD patients. Also, 232 (57.8%) of the respondents have difficulties in reading street signs and the name of stores, the non-SCD respondents were the most (65.5%). Among the respondents (68.2 %) 274 have no difficulties in doing their activities at night, 151 (55.1%) of them were SCD patients. Most of the respondents 332 (82.6%) have no difficulties in picking and matching their clothes, 72.5% (241) of them are SCD patients. 84 (20.9%) of SCD respondents have poor vision, in the other hand 108 (26.9%) of them are with good vision as shown in table 4

## DISCUSSION

This study was carried out because SCD has high prevalence in Saudi Arabia, especially in eastern region <sup>(6)</sup>, and the review of the literature suggest a causal link between retinopathy and SCD <sup>(10)</sup>.

The analysis of the results revealed that there is no significant difference between the 2 groups of the study in relation to the visual quality assessment.

Regarding the age, the age group >40 had the lowest visual assessment score. Which was expected and attributed to the undiagnosed age related eye diseases like presbyopia and subclinical cataract?

The data analysis surprisingly showed that there is no significant correlation between sickle cell disease and the quality of vision (P value > 0.05). The responses to the questionnaire between the 2 groups had only minor differences. Despite this, we can't be sure whether SCD has direct effect on quality of vision or not based on the questionnaire only.

Compared to other studies conducted with different study designs <sup>(8)(10)(11)</sup>, retinal involvement in sickle cell disease had slight effect on quality of vision compared to the normal groups which is against the results in this study. Overall,

the hypothesis needs to be approved by a prospective study with long-term follow-up.

Furthermore, the general vision, the near and distance activities, color vision and peripheral vision scores were the significant predicting factors for the vision-specific VFQ 25 questionnaire.

Factors affecting the general vision, the distance vision scores were the age and health status (sickler or non-sickler), these findings are comparable to other studies using the VFQ-25<sup>(12, 13)</sup>.

Health status as a factor doesn't predict the near activities, peripheral vision and color vision scores. We can attribute these results to the fact that peripapillary, macular arteriolar and retinal vein occlusions are rare with SCD<sup>(14, 15)</sup>

Non-response rates for activities such as newspaper reading, driving, and watching TV may attribute to the elderly age of the study respondents and their incapacity to do, or lack of desire to engage in these activities.

Two limitations were in our study. First, a cross-sectional type of study was used to obtain the data due to limited resources; consequently, we could not measure study respondents' long-term visual function and change in the quality of life over time. Second, an electronic survey was distributed. Therefore, we can't ensure the validity of some of the responses.

As a recommendation to the future researchers is to open the study population to different regional areas and to use prospective cohort study to help thoroughly understand long-term QOL changes over time for SCD patients.

## CONCLUSION

The study concluded that there is no significant relationship between sickle cell disease and vision disabilities. We recommend other types of studies to be conducted in this important field, especially in the eastern province of Saudi Arabia.

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